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The Surgeon and the Child

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Preface

In this book I have tried to tell the story of children's surgery for those who meet and must help solve the surgical problems of infancy and childhood. Because it is just as important to recognize a surgical problem as to treat it, much space has been given to consideration of diagnosis. Obstetricians, pediatricians and generalists are the first to see the deformed or surgically ill child and upon them rests the responsibility of securing adequate care at the proper time.

This is not a standard textbook of pediatric surgery. For the sake of brevity, embryology, references to medical literature, extensive illustrations and details of surgical technique have not been included. If credit were given to all who have contributed to pediatric surgery, each page would be liberally sprinkled with names, and Dr William E Ladd's name would appear in every chapter. Excellent standard textbooks on surgery of infants and children have been written by American, English and German authors and contain far more detailed basic informa

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tion than will be found in this book Those who wish to learn all that is known about any phase of pediatric surgery—any single operation—will still, after reading all available books, have to search through the great store of literature for details and differences of opinion Finally, the seeker of knowledge will have to add personal visits to the workshops of those who appear to know most about some particular subject

An attempt has been made to reflect the attitude of the surgical staff at the Children's Memorial Hospital of Chicago toward some of the surgical problems common to infants and children Emphasis has been focused upon certain points in diagnosis and specific methods of treatment, often in a somewhat dictatorial fashion These views derive in a large measure from personal experience—that painful process of trial and error—and from contact with many sources medical literature, suggestions of physicians, residents, interns, nurses, medical students and parents A little soul searching will reveal that most of one's accumulated information has been picked up from others bit by bit These fragments of knowledge true today and probably false tomorrow have been pieced together in simple form for easy reading

In keeping with the custom of dedicating a book to somebody, I want to dedicate this book to the infant who has the great misfortune of being born with a serious deformity All life is before him, and what is done during the first few days may decide whether life will be a joy or a burden If this infant could speak, it would beg imploringly of the surgeon, "Please exercise the greatest gentleness with my miniature tissues and try to correct the deformity at the first operation Give me blood and the proper amount of fluid and electrolytes, add plenty of oxygen to the anesthesia, and I will show you that I can tolerate a terrific amount of surgery You will be surprised at the speed of my recovery and I shall always be grateful to you"

WILLIS J POTTS

Chicago Illinois

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The Cry of Children

"With no language but a cry" children are asking for better surgical treatment of their ills and are begging for more thoughtful attention to the congenital deformities it was their misfortune to be born with The newborn infant has no words to demand his rights and is aided only by a couple of flustered parents dismayed at the sudden misfortune of an unanticipated catastrophe Many of the most severe ills especially of infants, are emergencies which preclude thoughtful analysis by parents of the surgical care their children are about to receive

During the past few decades the medical and surgical professions have turned a listening ear to these insistent cries and are now attempting to answer them with improved care Pediatric surgery born of the law fully wedded parents—need and demand—is now joining the family of specialties After slow progress during infancy the specialty has grown into a lusty child howling for recognition and demanding a place at the table. The pablum of increased knowledge has stimulated growth

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Twenty years ago the rare pediatric surgeon was looked upon as a peculiar person who for some reason known only to himself chose to operate on "little people" Need and demand have changed this viewpoint Physicians are seeking and parents are demanding that children receive the same expert surgical care which pediatricians offer for their medical ills

The field of pediatric surgery has broadened rapidly Previous to about 1940 surgery of children, except in a few hospitals, included primarily operations for pyloric stenosis, intussusception, hernia, appendicitis, osteomyelitis and empyema Since then have been added surgical correction of congenital anomalies of the esophagus, operations for congenital heart disease, knowledge of lung cysts and lobar emphysema, better understanding of the treatment of atresia of the rectum and associated fistulas, operations for Hirschsprung's disease and meconium ileus, tremendously improved preoperative, operative and postoperative care, and a host of others All operations on infants and children have improved spectacularly in recent years In fact, so much has recently been learned that the pediatric surgeon must run to stay where he is, and at the same time must struggle with innumerable problems still to be solved

Much uncertainty and confusion exist at present about the position of children's surgery in the company of the great specialties which have matured and occupy unassailable positions in the medical world There is little question, however, that during the next twenty-five years every leading medical school and large medical center will have a department of pediatric surgery, manned by a division head under the department of general surgery Such a chief of service will take his place alongside the chief of pediatrics and all other specialties Upon him and his associates will fall the task of giving children the surgical care they deserve

To meet the demand for better surgical care of children, more trained men are needed, and facilities for such training are slowly being provided Universities are beginning to recognize the desirability of having a children's hospital as part of their medical teaching program Such a hospital is best run as a separate and complete unit staffed by men who give most if not all of their time to all phases of child care Every activity of the hospital, from playroom to instruments in the operating room, is geared to the children's level In this atmosphere the trainee, grounded in the principles of general surgery, absorbs the atmosphere of total child care A half-floor for children in a general hospital is satisfactory for routine illnesses, but is not adequate for surgical training for the simple reason that surgical cases are too few The most difficult problems of children's surgery—severe congenital deformities—are rare In the average general hospital with an average obstetrical

service too few remediable deformities of the newborn occur to satisfy the requirements of training for a budding pediatric surgeon

More children's hospitals are needed In all centers of dense population children's hospitals should be strategically located To these centers ideally the difficult medical and surgical cases should be sent Present adequate and fast transportation makes it possible to move with safety almost any emergency case to a distance of up to 100 miles There are in this country approximately fifty children's hospitals well staffed by capable pediatricians but less than fifteen of them have adequate diversified surgical departments capable of rendering all types of surgical care It sometimes seems that the infant and child have been forgotten—not by the physician or pediatrician but by the surgeon Their surgical ailments are so complex that no one man is capable of restoring all deformed and sick children to good health and yet that remains our objective The infant is here it needs help and adequate surgery is essential

That the desired objective will eventually be reached is unquestioned because the rewards of children's surgery are so great The satisfaction of correcting a deformity in a newborn infant lies in the fact that all his life lies before him Parents hope for miracles but are grateful for the best that can be given by a mere human being Pediatric surgery is constantly fascinating and tremendously rewarding It is broad enough to interest the worker difficult enough to satisfy the ambitious and new enough to stimulate the imagination The infant "with no language but a cry" and the child with no words to express the desire to be well and normal ask that we make available to them the benefits of increased knowledge of their surgical diseases

The Deformed Child

Congenital anomalies of all kinds are relatively common The majority are easily corrected or soon forgotten Oddly shaped ears, a deformed hand a crooked leg, even a birthmark on the face—all are of little actual consequence and soon become part of one s personality Approximately 4 per cent of infants are born with extensive deformities some of them incompatible with life others partially or completely remediable by surgery A few infants—an estimated small fraction of 1 per cent—are born with such extensive and complicated abnormalities that surgeons can offer little more than lifesaving procedures This small group forms the basis of this discussion

Within a period of a few months three seriously deformed infants were admitted to the Children s Memorial Hospital All had somewhat similar defects, but each was attended with quite different circumstances

The first infant, a two-day-old son of farmers, was born with exstrophy of the bladder atresia of the rectum bilateral club feet, a Klippel Feil deformity of the neck and unquestionable microcephaly

It was explained to the father, who accompanied the child to the hospital, that some of the defects were in part remediable, but that no treatment could alter the size of the head and brain It was further explained that immediate operation upon the rectum was necessary as a lifesaving procedure The father's probing and intelligent questions about the likely future for the child were frankly answered Stoically and unemotionally, he simply said, "There will be no emergency operation." There were three normal children at home

The second case, a first-born infant of young parents nineteen and twenty years old, was a sort of half-child, the left arm, leg and ear were missing, and the left side of the head was so misshapen that the left eye protruded from an inadequate socket Besides these rather terrifying deformities, the infant had atresia of the ileum In spite of all these abnormalities, the infant seemed vigorous and cried lustily All four grandparents and the father came to the hospital with the child They were told that to save the baby's life release of the intestinal obstruction was necessary They then put this question, "What do you advise?" No more difficult question was ever asked, and my reply could only be, "I can't answer your question You talk it over and let me know your decision Whatever you decide, I'm sure will be right" It was impossible not to add, "The parents are young, and the chances of having a number of healthy children are excellent"

The relatives, devout members of a Protestant faith, requested the office for private discussion and prayer Upon my return they said that they had decided the baby should have a chance to live Operation was performed, and the baby survived Was their decision wise? A few days later one of the grandmothers said to me in the corridor, "I wish God would take it"

The third case was that of an infant born with an omphalocele, exstrophy of the bladder and a cloaca so extensive that the entire perineum was open Sex determination was impossible Besides these obvious deformities of the bowel, there was duodenal obstruction most likely due to malrotation of the bowel The relatives were advised that only urgent surgery could save the baby's life, that many later operations would be necessary, but that no amount of surgery could give them a normal child The parents were Catholic and called in their parish priest for advice He told them that under unusual circumstances in which extraordinary care is needed, extraordinary skill required, extraordinary expense involved and extraordinary suffering incurred, it is not a sin to withhold treatment Although there was little question that the priest's advice represented the opinion of the Catholic church, authorities on this subject were consulted by the author

Edwin J Healy, S J , in a book on "Medical Ethics," published by the Loyola University Press of Chicago in 1956 and approved by the late

Cardinal Samuel Stritch states "The principle which serves as a guide in regard to the use of extraordinary means is this *one may but need not use extraordinary means to preserve life* Obviously the problem is to distinguish clearly between what may properly be considered extra ordinary and what is ordinary We may define as an extraordinary means whatever here and now is very *costly* or very *unusual* or very *painful* or very *difficult* or very *dangerous* or if the good effects which can be expected from its use are not proportionate to the difficulty and inconvenience that are entailed" No operation was performed and the child died.

Jewish Protestant and Unitarian churches have no written opinions which may serve as a guide to parents under such circumstances

In the first case the father a practical farmer came to a quick decision In the second case the relatives chose to give the baby a chance to live In the third case any feeling of guilt which the parents might later experience because of refusing operation was avoided by the established opinion of a church

Unusual circumstances surround each such case as it appears and render past decisions meaningless

A number of years ago a middle-aged couple brought in their only child a six year old idiot son deeply cyanotic because of tetralogy of Fallot The child could not walk more than fifty yards without collapsing. He was unable to feed himself and responded to emotional stimuli only with a sort of guttural grunt Although the parents understood that improved oxygenation would not benefit their child's mental condition they urgently requested operation Six months after a successful aortic pulmonary anastomosis when the parents returned with their boy for routine follow up examination they grasped my hand and with tears of gratitude in their eyes thanked me for helping their boy The mother proudly said, "Now M can walk all the way to the post office and back without stopping"

It is difficult to measure devotion or to sound the depths of love for a living thing Those who have been fortunate enough to have been spared the tragedy of a seriously deformed child are incapable of guiding or advising the unfortunate An infallible answer to the question of performing or withholding lifesaving operation for extensively deformed infants is not always available. Each case differs slightly from those seen before it may be the only child or it may be the last in a family of seven Religious beliefs philosophies of life and economic conditions alter decisions This fact is unquestionable When in circumstances as outlined above, parents make a choice, the surgeon must support their decision, if it is against operation, they should be supported lest they later are plagued by a feeling of guilt, if in favor of operation, every

thing possible must be done to save the hapless child and make life as tolerable as possible

Life is the most precious thing in the world Is it so to the individual born without a brain capable of comprehending existence? Is life worth while when burdened with deformities that make social seclusion necessary and marital love impossible? Maybe it is Is it fair to demand that for a deformed and idiot child parents spend their savings for hospitalization, operations, nurses, blood transfusions, oxygen and drugs while they deny to their other normal and intelligent children advantages to which they are entitled?

Our duty as doctors is crystal-clear—to preserve life And yet, when looking at a hopeless little mass of deformed humanity, compassion wrestles with duty

The Heart of a Child*

The physical heart of a child is just a piece of living muscle marvel ously adapted to its sole function of pumping blood It is a rugged mechanism that will tolerate the ravages of infection the scars result ing from impaired blood supply and the approaches of surgeons tools It is the most efficient self sustaining pump in the world In a philosophical sense the heart of a child is a delicate mechanism, sensitive to the slightest wounds of fear insecurity indifference thoughtlessness and misunderstanding Many centuries before its physical function was known the heart was considered the seat of all emotions and impulses, good and bad From earliest infancy the heart of a child is subject to a constant stream of mental and physical stimuli, but for the purpose of this discourse it reflects the results of stimuli with which it is bombarded during those short phases of life when the infant or child suddenly meets head-on with doctors illness, hospitalization and operation.

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Child-Doctor Relationship

Little children are afraid of doctors because of previous unpleasant procedures to which they have been subjected They use no system of logic in evaluating the virtues of medical or surgical treatment Primarily they fear needles and "shots" For children's protection physicians have to do unpleasant things to them, such as giving painful immunizing injections and administering antibiotic drugs Consequently, when taken to a doctor, the child promptly bursts forth with, "Are you going to give me a shot?" A child associates the prick of a needle with a white coat or a nurse's uniform We might as well admit it—until we have completely won the confidence of children we are ogres to them Children are in a sense like dogs—they instinctively recognize antagonism and intolerance It is a truism worthy of repetition that a doctor's successful approach to a child is based on a fundamental love of children and a cultivated tolerance of their eccentricities However, in defense of the doctor, I believe that the screaming, not too ill, uncooperative child, hovered over by indulgent parents carrying on unintelligible baby talk in a vain effort to comfort their little darling, is a trial of patience to any doctor A carefully developed deaf ear to unjustified screaming, a zealously masked look covering signs of irritation, and a completely camouflaged desire to stuff one's handkerchief in a wide open mouth pay dividends A little time and even a smile, if authentic, will soon bring peace and cooperation Interestingly enough, many of the "badly behaved" children become very amenable when they find out that after all the doctor is their friend and is actually trying to help them

The child reflects the attitude of its parents The mother who constantly worries about her child's every tiny variation from normal—what may not even be termed an illness—will soon have a child who magnifies complaints and courts disease The seed of psychosomatic ailments is planted and will bear much fruit in later life Mothers in general know that about 90 per cent of children's ailments are self-limited, but anxiety about a possible serious illness often overshadows good judgment Parents neglect their own infirmities, but when their baby is sick—especially the first one—they fly to the doctor That is good for the pediatrician, but not for the child *It too soon learns the power of accentuating* complaints and thereby obtaining that delightful status of holding the center of the stage

Actually, there are few diseases that will not allow a reasonable time for intelligent observation by parents Those few diseases that require prompt medical attention have such specific and characteristic signs of variation from normal that an observant mother may promptly recognize the need for immediate help For most of the simple ailments, not severe enough to keep a child away from his favorite television pro-

gram it makes little difference whether a tablet of aspirin is given or a bit of goose grease is rubbed on the sore place

Impact of Hospitalization

The doctor's primary concern is with the effect upon the emotions of the infant or child whose illness is severe enough to require hospitalization and operation I have often wondered what sort of scar how deep and how serious is left on the heart of a child who is torn from his parents and suddenly tossed into hospital environment associated in his mind with insecurity and pain Because people get sick there must be hospitals Adults are presumably reasonable creatures who when ill go to hospitals more or less willingly because they know they have to do so to get well Doctors work in hospitals every day and think them wonderful but as patients most of them hate hospitals and their routine Imagine how a child feels Children's hospitals are marvelous institutions to parents the day they take home their child who was ill and now is well That does not change the fact that to a child a hospital experience is often a nightmare Before the age of reason a child is unable to comprehend why he should have been subjected to the emotional insecurity of separation from his mother Even the finest hospital falls far short of what an ideal hospital should be If nursing service were limitless and if laws of economics could be repealed there would be no problem A mother then would simply bring her child to the hospital and deliver it to a nurse well trained in scientific care of children and in the art of substitute motherhood. Whether the child remained in the institution three days or three months would make little difference Normal development would not be impeded and emotional turmoil would be avoided It could be as simple as that

Of necessity hospital care for children has become extremely complicated and, consequently has a tendency to become too impersonal We order intravenously given fluids blood transfusions injections of drugs, nasal tubes blood cell counts and a host of other things All are necessary for recovery of the patient, but how can a little child comprehend this, and how can residents nurses and technicians get their work done if they have to spend two thirds of their time explaining to unwilling listeners the object of each move? When dealing with immature and uncomprehending minds parents and doctors have to do the best they can to minimize the rips and tears in the emotional patterns of the children Little is known about the memory of a child during its first year of life but it is known that reactions to fear and insecurity manifest themselves early An infant up to about one year of age is indifferent to physical surroundings but not by any means indifferent to the people who care for him Ordinarily it is believed that the infant aged six

months or less does not care whether the mother is present Actually, the infant does not care so long as a substitute mother gives the same tender loving care Nurses act as substitute mothers Because they are women, and in their not too subconscious minds are looking forward to the day when they will be caring for their own children, they easily learn the technique of administering tender loving care to the young infant Whenever possible, all infants are fed by a nurse holding the baby on her lap Sick or well, the infant loves contact Observe in unspoiled nature how a mother animal guards, fondles and cares for its young, and you will realize the utter folly of the advice to "leave the child alone" An animal outgrows babyhood rapidly, but the human being, helpless for years, requires long and constant security provided by mother The child needs protection and love given by instinct, uninhibited by theoretical advisors and professional baby raisers

Understanding the Sick Child

Infants require stimulation—auditory, visual and tactile Their development—entirely by the route of their senses—must continue during those periods of time that are spent in a hospital Of course, an illness of a week or two is going to leave no scar The child's memory of pain is short, and the ability to harbor resentment has not yet developed Prolonged illness is another story It has been shown again and again by psychiatrists that infants do poorly in a foundling home where they get a minimum of attention Even under the most hygienic surroundings they develop poorly, physically and mentally The mortality rate is higher in these children than in even a rather poor home where they get essential tender loving care This case came to my attention recently A six-week-old normal male infant was brought to the hospital with a broken femur It was necessary to put both legs up in overhead traction The infant lay on its back twenty-four hours each day and was fed in that position Circumstances were such that he had no visitors After about three weeks the infant lay in his bed in a constantly listless state, indifferent to food and people He did not cry or fuss—just lay there with eyes half closed and, one might say, shut off from the world At this time a volunteer worker was assigned to this infant eight hours a day She fed the baby while supporting his head and shoulders During waking hours she played with and fondled him In another three weeks, when ready to go home, he had gained weight and was smiling, cooing, and acting as a three-month-old child should This infant had not been neglected He had been fed, bathed and kept clean, but that is not enough for any child, sick or well They need thoughtful stimulation, and that stimulation of their senses is what gives security and pleasure in living, synonymous with growth and development

An eight month old baby girl so severely burned that recovery seemed impossible was kept alive and recovered because of nursing care Plasma could not be administered fast enough to replace proteins lost from oozing wounds Only food could keep up the child s strength It is not easy to get food into a child who has constant pain and a tempera ture of 103 to 104° F (39 4° to 40 C) every day It was accomplished by patience As soon as the child could be picked up she was fed on a nurse s lap night and day She recovered She has physical but I am quite sure no emotional scars When the mother took her child home she thought a facetious suggestion that she include in the child s name the initials R. N had been seriously made

To mothers I suggest that the child be given the attention he craves sick or well A sick baby in the hospital should be visited every day and at the earliest possible date he should be taken home There is no place in the world like home for a child Even the poorest home where there is accord is better than the finest hospital If the child must remain in the hospital long visit often and crowd in as much attention as possible during those few hours Whatever spoiling may be done during visiting hours will be counteracted during the rest of the day and night. The child will naturally cry when the parent leaves but return visits will dispel the fear of being forgotten or deserted

The sensitive three-year-old or four year-old children requiring hospitalization and operation are the ones for whom I am sorry While seriously ill they have not enough energy to scream for their parents but the moment they improve they begin "hollering" for their "mommy " Many many times I have seen such little children standing in their high sided cribs their faces stained with tears and their pupils dilated with fear sobbing interminably "I want my mommy " You can pick them up offer them toys and tell them over and over that "mommy" is coming. Their only response is more wailing and "I want my mommy now " A three year-old child has no sense of time "Pretty soon" means absolutely nothing. Anything he wants he wants right now A three year-old boy had a rather simple operation normally requiring about a five-day stay in the hospital. He was in a ward—no private room was available When the parents left after visiting hours he began to scream and continued most of the night and the following day until the parents returned There was nothing to do but let the child go home No physical complications could be worse than having this youngster tear his emotional system to pieces

Children from five to eight years of age are often equally unhappy but can be steered into a more equable frame of mind during the long twenty-one hours between visiting periods, provided there are enough nurses and helpers to give them their time and personal attention In security and fear to a far greater degree than is realized, plague these

months or less does not care whether the mother is present Actually the infant does not care so long as a substitute mother gives the same tender loving care Nurses act as substitute mothers Because they are women and in their not too subconscious minds are looking forward to the day when they will be caring for their own children, they easily learn the technique of administering tender loving care to the young infant Whenever possible, all infants are fed by a nurse holding the baby on her lap Sick or well the infant loves contact Observe in unspoiled nature how a mother animal guards, fondles and cares for its young and you will realize the utter folly of the advice to 'leave the child alone An animal outgrows babyhood rapidly but the human being, helpless for years requires long and constant security provided by mother The child needs protection and love given by instinct uninhibited by theoretical advisors and professional baby raisers

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pick the area from which such a patient comes The percentage of correct guesses eliminates chance as a factor Children who come from small towns especially from farms can be spotted in a moment They are easy to care for accept hospital routine and do not scream when being examined Say to one of these children "May I listen to your heart?" and the child lifts up his gown and calmly accepts half a dozen stethoscopes on his chest Why is this true—if it is? I believe these children are so because they come from stable closely knit families Many come from farms where the family is the center of all activity These children know from experience and few words of direction what they may or may not do When their parents tell them they are to go to a hospital and must be good boys or girls they are just that It is routine for them to accept what comes from day to day The evening before operation parents are naturally on edge If they were not concerned they would not be quite normal Outside the door they ask about the danger of the operation and they get what they are entitled to—a frank answer The truth of the matter is that today with good anesthesia guarded asepsis antibiotic drugs and trained surgeons most operations are safer than a cross-country trip in an automobile

Preparation for Operation

At Children s Memorial Hospital the child is told exactly what is going to happen Often the youngster will ask, "Are you going to cut me?" A bit of license is taken at that point, and he is told "We are going to operate upon you and fix that sore place" If the child is three to five years old, he is told that in the morning he will be moved in his bed to a special room where he will see nurses and doctors with funny things called masks on their faces He will be put on an extra fancy table and be allowed to blow up a balloon As he blows up the balloon he will get sleepy and when he wakes up he will be back in his room where mommy and daddy will be waiting for him. It is as simple as that The older child, six to seven years of age is likewise told in words that he can understand exactly what will happen After the questions have been answered the child is content and goes to sleep The mother is the one who gets the sleeping pill One child said after the explanation about anesthesia and operation "You don t have to tell me all that stuff I know all about it—I saw it on TV"

I began to explain to an eleven year-old girl, on the day before her operation for a patent ductus arteriosus what was expected to be done on the following day The mother jumped up grabbed me by the arm and steered me out of the room "Nancy doesn t know what she s here for I don t want her to know She thinks she s here for another test." I promptly led the mother back into the room and said to Nancy "Do

children who are sick and, against their wishes, are forced into a hospital The best that can be done in the present state of hospital tradition and medical necessity is to prepare them for the unpleasant incidents that suddenly face them and in love and understanding soften the blow to their immature minds For older children nothing in the field of medicine is finer or more essential than an understanding nurse, resident or intern who, with cultivated sixth sense, makes them feel that they have not entered torture chambers, but have been admitted to a place where folks are going to help them get well and where they are interested in their happiness

Child's Reaction to Illness and Hospitalization

It never ceases to be interesting to watch the reaction of children entering a hospital for operation It varies all the way from childish bravado to sheer panic A seven-year-old boy was brought in because of a question of hernia Examination proved there was no hernia, and as the boy left the examining room he made a gesture of wiping sweat from his brow and exclaimed, "Boy, that was a close one" A younger child clarified his position after I had explained to him that he would have to have an operation He said, "I hate you, you stinker" Another little boy said in response to what he considered bad news, "You know what? Lions eat people, and I hope they eat you" One will not have to *worry that such children will have repressions*

Children growing up in an atmosphere of freedom will be well adjusted extroverts if these liberties are balanced with intelligent restrictions Some children, unfortunately subjected to no limitations at home, expect the same indulgences when entering a hospital No attempt is made to alter a child's social pattern during a brief hospital stay, but staff members are forced, for the good of all, to impose some limitations upon children who are patients for many months It has been interesting to observe that children actually enjoy the intelligent restrictions of an orderly life Children love established routine When they learn what they may do and what they may not do, they become cooperative, are happier, and are more secure than when they are completely unrestricted

Hospital personnel must deal with every type of child from the overindulged child of supersolicitous parents who expect every whim to be granted upon demand to the affable child who patiently accepts without objection the routine of the hospital Children, like some adults, learn rapidly that the gentle voice and the affable smile get them much more service than harsh and sullen demands An interesting observation has been made at Children's Memorial Hospital on the adjustability of children Patients to be operated upon for congenital heart disease come from many localities and many states It has become a sort of game to

Preoperative Care

Physical Examination

Preoperative care begins when a person decides to study medicine During the long period of training attentive and enthusiastic observation of all manifestations of disease from its earliest symptom to final tissue examination at the postmortem table contributes to the art and science of evaluating a patient's condition Those who enter the field of pediatric surgery finally bring into high power focus their attention upon the surgical diseases of children With no history from the infant or small child except the observations of distraught parents and with only a distorted history from the older child, who may be annoyed at being brought to the hospital, the pediatric surgeon is faced with the necessity of deciding, on the basis of physical findings alone the type and degree of illness

THE INFANT In this book the interchangeable words "infant" and "baby" mean a child below two years of age

The sick infant is seen by the pediatric surgeon at the hospital in

you know why you are here?" "Sure," she said, "I'm going to have an operation on my heart." You cannot fool children, and, if you try, be prepared for a terrific backfire. When Nancy is sixteen she will be fooling her mother and be an expert at it because of her early first-class tutoring.

Children get such distorted ideas of hospitals and operations. A six-year-old girl was admitted to the hospital for an operation. When I entered her room, the child was clinging to her mother and sobbing hysterically. Nothing would comfort her. After long questioning about the usual things that cause pain she finally blurted out, "I don't want to die." Then the full story came out. A few weeks previously one of the children from her room in school had been taken to a hospital for operation and had died. It is easy to understand how her immature mind, knowing nothing about the ailment of her playmate, jumped to the conclusion that all children admitted to a hospital die. She was finally persuaded to go on rounds through the hospital to see other children who had had the same operation she was to have. I shall never forget that little, cold, moist hand in mine as we visited other convalescent patients. One could see the cloud of fear being dispelled as she stared at the children in their beds coloring pictures or playing with toys. She returned to her mother with a smile. Incidentally, she went through the operation and postoperative period without a whimper.

An eleven-year-old boy who had been in the hospital six months and had had seven operations and more than twenty blood transfusions said to his surgeon when he finally recovered and was about to leave the hospital, "Even after all those operations I still like you." Children are such amazing little creatures. Tell them in simple words why they have to go to the doctor or the hospital or why they have to have an operation, and, in most instances, they will cooperate in a fashion that adults might well emulate. Faith and trust are completely unspoiled when children are dealt with honestly. So little effort, so great the reward.

Comment

The mystical heart of a child is a precious and beautiful thing. It is marred only by wounds of a thoughtless and not too intelligent world. In a physical sense the heart is a tough organ, a marvelous mechanism that, mostly without repairs, will give valiant pumping service up to a hundred years. In an emotional sense it is susceptible to wounds of indifference, thoughtlessness and neglect, and during episodes of illness is especially vulnerable. The heart of a child is mysteriously molded by parents, teachers, playmates and all those with whom it comes in contact. Physicians wish during those short but violent episodes of illness to avoid wounds that will leave irreparable scars. I am convinced that the heart of a child sunned by love, security and understanding will be able to withstand the storms of illness and pain.

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the examining room. Usually the family physician or pediatrician has already examined the baby at home and decided that hospitalization is necessary, or the infant recently born in a general hospital is transferred for care to a children's hospital. Regardless of whether the child has been previously examined, it is well for the surgeon to re-examine and thereby keep his diagnostic armor shiny.

The baby is laid on the examining table, and all his clothes—diaper, booties, everything—are removed. The room must be warm, not only to protect the infant, but also to avoid criticism by the mother. A glance will tell much about the baby's condition. The parents will have called attention to some particular area as the source of trouble, but for the moment that is disregarded in favor of general observation.

Although it is a well established routine to weigh all patients upon admission to the hospital, the preliminary examination concerns itself only with the general appearance of good or poor development commensurate with age, and a history of rapid or slow weight loss.

If a child is over six weeks of age, a few moments spent in trying to make the baby smile is time well spent. The mother is pleased when the baby smiles and delighted that the examiner shows personal interest in her wonderful baby; the doctor sees the smile and knows, first, that the baby is probably not acutely ill, and, second, that he probably is developing normally. If he doesn't smile, he may not be sick, but just indifferent to the doctor's variety of clucking. If the baby's age is three months or more and he has never smiled, that is an ominous sign.

Perhaps the baby has not recently been seen by the family physician or pediatrician and the mother has not been told or become suspicious that the child is seriously ill. Slanted eyes, loose joints and general flaccidity at once suggest mongolism. Further questioning should be guarded or completely avoided. The pediatric surgeon seeing the baby for the first time should not reveal to the mother his suspicions, no matter how right he may be. It is far better to suggest in an offhand manner that there is a possibility that the baby is not developing altogether according to schedule and that it would be wise later to consult her doctor. The heart of any mother will be broken when she learns that her baby is not normal, but let her own trusted physician as gently as possible deliver the blow.

The baby who cries lustily frightens the parents more than the doctor. So long as the baby can yell, the outlook is not bad. The sick infant who merely whimpers and looks sick is in acute danger. Scores of similar observations are made in less time than it takes to write these sentences.

Look for beginning rashes. If any suspicion presents itself, look for Koplik spots in the mouth at once and feel for enlarged glands behind the ears. Nothing is more embarrassing than to admit a patient with

fever and vomiting who on the following day breaks out with measles Be careful! That tumor over the parotid region may be ordinary mumps What's mortifying is the elevation of the pediatric staff's collective eye brows when a surgeon admits a patient to the hospital with some contagious disease

It requires only a moment to raise the head and eliminate the possi bility of stiff neck due to meningitis

A pinched face sunken and lusterless eyes red lips a dry tongue and red palms and soles write out the story of fever and dehydration A fleck of green vomitus at the side of the mouth makes one suspicious in an instant of intestinal obstruction The baby who refuses a nipple and restlessly moves his head from side to side and makes motions with his tongue as though trying to spit something out is as certainly nause ated as if he said "I feel like vomiting"

Rapid breathing unequal expansion of the lungs suprasternal re tractions crowing respiration—such findings together or separately signal a serious defect of the respiratory system

Cyanosis without rapid respiration suggests congenital heart disease or birth injury to the brain cyanosis with labored respiration calls for investigation of the respiratory passages from pharynx to bronchi for intrinsic or extrinsic obstruction and for consideration of such conditions as atelectasis diaphragmatic hernia, lung cyst and pneumothorax

It's a fascinating game to try to decide first of all whether or not the baby is seriously ill and then from observation alone and from the history given by the mother to make a tentative diagnosis Never under estimate the mother's observations if the baby being examined is other than the first born

The routine physical examination made by hand stethoscope tongue depressor and otoscope is so well covered by our pediatric confreres that little need be said here

Most diseases of infants requiring additional information obtainable by palpation are confined to the abdominal region With a warm hand—no speck of dirt beneath the fingernails—the abdomen is examined first by a gently sweeping motion to prevent muscle spasm Distended loops of bowel and masses not visible can best be felt by superficial palpation Only after the abdominal wall has become accustomed to the examining hand can one find a pyloric tumor an intussusception, a kidney tumor a cyst or an unsuspected localized abscess A pacifier covered with sugar will often distract the baby's attention and allow satisfactory examination The baby who cries continuously and is too sick or too annoyed to accept a nipple presents a problem in patience Usually it is wise to try again later after a bit of sedation has been given No abdominal examination is complete without palpating the inguinal areas for hernia and for femoral pulse beats.

Rectal examination is done as indicated, but rarely yields much information unless the history and symptoms suggest rectal disease or a lesion in the lower part of the abdomen

Even the surgeon looks at the throat in every child who has a fever, and unless the pediatrician has just pronounced the ear drums normal he also uses an otoscope If there are obscure symptoms of appendicitis, if the temperature is high and the abdomen has yielding rigidity and obscure generalized tenderness, pneumonia is suspected and not ruled out until an x-ray picture of the chest has been taken

A history of long-standing, intermittent, unlocalized abdominal distress, poor feeding and failure to gain weight calls for an excretory pyelogram A rounded mass in the suprapubic region is a distended bladder

A tentative, and not infrequently a definitive, diagnosis is the reward of patiently listening to the mother's story, critically observing the infant and carefully doing a methodical examination At least one will have decided that the baby is or is not seriously ill, and does or does not require further study and hospitalization

THE OLDER CHILD Examination of the child from two to six years of age requires an altogether different approach These children are little animals who have not reached the age of reason and who react to conditioned stimuli The majority are affable, cooperative and easily manageable, a few are little demons, attractive in their stubbornness, but determined to fight an examination to the last ditch All who have had some medical experience fear the doctor and invariably ask as they enter the door, "Are you going to give me a shot?" They hate needles After assurance (by a showing of open hands) that they will receive no shot they hesitatingly check off that danger Still they are suspicious, however, of strange surroundings and have to be convinced that they may trust a doctor never seen before

All children love attention Every child has something attractive which may be admired Establishing rapport with a child is simply a matter of salesmanship If a salesman can get a prospective customer to say "yes" to a sufficient number of questions, eventually the customer will say "yes" to the purchase of something he actually doesn't want "My, what a pretty dress Is it new?" It always is "Do you like cowboy suits?" "Are you a good little girl?" I've never had a negative answer to that one One can go on with any number of questions Slowly the ice melts, and finally after hitting on something that evokes enthusiastic attention one motions to the child to come As soon as the child of its own volition walks to the doctor, the battle is won and the examination is on its way But woe unto him who breaks faith in promising that nothing bad is going to happen and then hurts the child I won the confidence of a fire-eating, six-year-old boy and during the examination thoughtlessly did something that hurt him He let off with a flow of language

worthy of a sailor's parrot He lisped on the last of the expletives that sounded like "bith" but there was no mistaking the music It was priceless but I am sure the father was sternly reprimanded that evening for being a bit loose with his outbursts about the house

Completely disrobing the young child is often unnecessary and almost invariably objected to violently Little boys always want to keep their pants on They probably feel that so long as they have their pants at least partially on they can manage a get away in a real emergency Invariably boys while being examined for hernia keep pulling up their pants Children will allow examination of the chest and abdomen without hesitation so long as part of their clothes is left on Some youngsters develop an early sense of modesty which should be respected Although the child is usually not completely disrobed because examination is always pointed to some particular area, it is well in piece-meal fashion if necessary to do a thorough physical examination It is humiliating later to find that one has missed an unsuspected undescended testicle in a child examined for pectus excavatum or to have overlooked enlarged glands in the neck in a child suspected of having appendicitis

The pediatrician who examines the abdomen of a child brought in with a running nose may eventually receive for the extra effort the well earned reward of finding an unsuspected abdominal mass It is amazing how many Wilms tumors and abdominal neuroblastomas have been found by the careful pediatrician who extends his examination beyond the area of complaint

Examination of a child in this age group requires a bit of finesse if the complaint is abdominal pain The child is suspicious that he is going to be hurt and invariably tightens his abdominal muscles The examiner carefully avoids the tender area and with a light sweeping motion accustoms the abdomen to the palpating hand. Slowly after spending considerable time on the nontender areas he keeps repeating "Now you tell me if I touch that sore place" The child will make it clear by wincing crying or comment when the sore place is touched but will not be resentful because of the warning. With the aid of a little distracting conversation about birthdays or Christmas and by superficial rather than deep pressure the tender area is located or the mass outlined To the surprise and delight of the child suddenly the examination has been completed. A diagnosis has been made and a friend won

Hospitalization

Parents are often dismayed when told that their child requires hospitalization Concern is a little less acute however for those who have been foresighted enough to have purchased Blue Cross hospital insurance Parents want to know whether their child should go to a ward or a private room. Infants are routinely placed in the nursery Whether the

child goes to a ward or a private room is largely determined by family finances, the number of children in the family, the type of illness and the degree of dependency of the child upon his mother Needless to say, the child, unhappy with any kind of hospitalization, is included in the conversation and reassured that nothing terrible is going to happen to him It is explained that it will be fun to be in a ward with other children Children are gregarious and respond favorably to herd instinct Parents are apt to think that the noise and confusion in a ward will disturb their child Nonsense! Children sleep through all sorts of disturbances and are rather indifferent to the discomfort of others A child in a four-bed ward gets more attention from the nursing staff than the child in a private room Theoretically, the nurse has to go into a four-bed ward four times as often as into a private room Automatically, she observes each child as she enters the ward Unless a seriously ill child in a private room is constantly attended by parent or private nurse, ward service is safer

ASSIGNMENT TO SERVICE. Not so many years ago it was customary to assign every child admitted to hospital to the pediatric service The pediatrician made the diagnosis, decided whether and when operation was necessary and directed all the postoperative care Since pediatric surgery has expanded, and trained surgical residents devoting all their time to the care of children are available, it is appropriate to admit infants and children directly to the surgical service Consultations by the pediatric staff are frequently sought In fact, we have a fine custom at our hospital which promotes good rather than ill will, members of the attending pediatric staff and medical residents are free to see semi-private and ward surgical patients any time they wish Suggestions are always welcome, but they had better be good Residents representing different services are competitive That's good Friendly rivalry promotes good care and lessens errors The resident who crows a bit too lustily when right about some disputed matter of diagnosis or treatment will in time have to listen to the same music from the other side of the fence

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Fluid and Electrolyte Requirements

A newborn infant requires no sodium chloride the first day of life and very little fluid Thereafter he will need about 50 cc of physiologic saline solution and enough 5 per cent glucose in water to bring the total intake to 50 cc per pound of body weight for 24 hours From infancy

through childhood fluid requirements slowly diminish for example a 6-pound infant needs 300 to 350 cc., and a 60-pound child 1500 cc., in 24 hours The dehydrated infant or child will require twice these amounts

Solutions have been perfected to combat dehydration and electrolyte imbalance all are physiological and each has been devised to meet slightly different conditions Dehydration as seen in surgical patients and caused by excessive vomiting or necessary withholding of fluid is satisfactorily treated in the vast majority of instances by physiologic saline solution and 5 per cent glucose in water given separately or combined Additions of ammonium or potassium chloride or more concentrated solutions of sodium chloride are made only when blood chemistry determinations indicate deficiencies

The dehydrated infant or child admitted to hospital on the surgical service because of vomiting usually due to abdominal pathology should be given glucose and physiologic salt solution immediately Drawing blood for chemical determinations is in order but awaiting the laboratory reports before beginning treatment is not only unscientific but also stupid The child needs replacement of fluid and electrolytes and needs them right now and in generous quantities

There are three ways of giving a vomiting dehydrated child fluid but only one is really effective Proctoclysis for little children is absolutely silly When the procedure is ordered the nurses notes invariably read as follows "Proctoclysis begun Proctoclysis stopped Bed linen changed" Subcutaneous fluid may be given if dehydration is not severe or if at the moment no one is available who is capable of inserting a needle into a tiny vein Absorption is too slow however to be effective Intravenous administration of fluid remains the only sure and effective method If the child is likely to be operated upon within twenty four hours it is well immediately to do a "cut down" at the ankle and insert a polyethylene tube into the saphenous vein The "cut down" is done in the dressing room under local anesthesia The infant or small child will have to be wrapped in a restraining sheet and the leg will have to be fixed with adhesive tape to a small board well padded with sponge rubber to avoid pressure necrosis The procedure an ordeal for the child is unavoidable If operation is to be postponed, it is preferable to give fluid with a tiny needle in a scalp or hand vein and preserve the saphenous vein for use during operation and for the critical few days following operation In spite of all precautions a polyethylene tube in the saphenous vein is rarely effective more than three days either the tube becomes obstructed or thrombophlebitis requires its removal

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has not concentrated on milliequivalents, it is often good judgment for him to look wise and say little as his resident calculates the electrolyte needs of the child

Blood transfusion is a life-giving measure for the ill and debilitated child, a lifesaving measure during and after operation No major procedure should even be attempted without having a well functioning tube in a vein and sufficient blood in the operating room to meet any emergency Occasionally it takes more time to place a polyethylene tube in a vein in a premature infant than to do the operation itself The temptation to proceed with the operation without a secure, tested needle or tube in a vein must be curbed A 5-pound infant may not require more than 50 ml of blood, but that amount is the difference between success and failure

It seems incredible that only thirty years ago giving a blood transfusion was a real event Whole blood was still considered preferable to citrated blood and was given only in the greatest emergency With multiple syringes blood was drawn from the donor by one man while another firmly held the venous cannula in place with a hemostat A third man injected the blood into the recipient while his assistant held that cannula in place A nurse received the empty syringes, washed them and handed them back to the first man Five people to give a blood transfusion! And often the syringe got stuck and less than the intended amount was given In describing to the modern resident this method of giving a transfusion in the old days, he gets a look on his face which suggests, "How could you have been so stupid?"

Respiratory Infections

Any upper respiratory tract infection—sore throat, running nose or cough—if detected, precludes admission to hospital, if appearing after admission, it automatically cancels operation It is trying and costly for parents who have brought their child a long distance for elective surgery to be sent home and asked to return later It seems that children of early school age are in some phase of an upper respiratory tract infection about 25 per cent of the time In a children's hospital colds keep the operating schedule in a constant state of turmoil, and there is nothing one can do about the disrupted schedule, or the cold, but be patient and make later appointments In case an emergency operation is necessary in the presence of an acute upper respiratory tract infection, the danger of delaying operation is balanced against the danger of pulmonary complications

Vitamins

In moderate doses vitamins do no harm and in some cases are beneficial Vitamin K is given parenterally before and after operation to

all jaundiced children and those with bleeding tendencies. To promote wound healing infants routinely receive a daily dose of a combination of vitamins A, B C and D before and after operation If the child is unable to take medication by mouth ascorbic acid is added to the intra venous solutions

Enemas

These are hateful to children and are not given preoperatively except as required for rectal and bowel surgery Children with normal bowel innervation have fecal impactions less commonly than adults and don t complain of being full of poison if their bowels don t move every day A dose of mineral oil or a teaspoonful or two of milk of magnesia is usually effective and far less objectionable than an enema. The fact that enemas and enemies are often confused is more than a matter of mispronuncia tion.

Anesthesia

Never having administered general anesthesia to a child, I feel completely uninhibited in discussing this subject. My position is similar to that of the music critic who can t play a note yet listens intelligently to weekly symphonies and writes more or less discerning reviews The music critic recognizes fine music as he listens the surgeon appreciates skilled anesthesia as he operates.

Psychic preparation for operation is supported by proper preoperative medication. The occasional apprehensive child sleeps better if assisted by a suitable dose of Nembutal the night before operation Ordinarily children don t worry about the future and usually need no help to go to sleep If the operation is to be delayed until afternoon some Nembutal in the morning, especially for little children accustomed to eating and drinking frequently will lessen anxiety and make more tolerable a forenoon of abstinence

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respiratory depression except in one child for whom 1/16 grain of morphine had been ordered The nurse in calculating the dose had some trouble with fractions and figured that two 1/8 grains equalled 1/16 Artificial respiration was necessary for the child and sedatives were needed by the nurse after her interview in the front office. Use of the metric system will lessen such errors provided the nurse has had no trouble with the decimal system

Even though the child has been psychically prepared for operation by parents or physician he still will need some reassurance as he is laid on the operating table At the last moment he will lose his bravado or complaisance and think up all sorts of reasons why application of the mask or whatever contrivance is used should be delayed a bit longer Patient indulgence with the child who is old enough to understand will calm fears and finally restore the feeling of assurance that somehow the ordeal can be tolerated and might even be interesting

Selection of Agents

Induction of anesthesia is accomplished by various anesthetic agents A few whiffs of ethylchloride Vinethane nitrous oxide or cyclopropane will quickly produce enough unconsciousness to make the child unaware of the displeasing odor and choking sensation of ether Whenever possible, it is desirable to avoid forcible restraint of the child during induction A nurse or attendant holds the child's hands with a grip of reassurance rather than of restraint. If the child becomes unruly consciously or unconsciously during the first plane of anesthesia, a second person may have to help hold the child to keep him from bouncing off the table Mechanical hand or leg restraints are never used.

Simplicity is the keynote of good anesthesia. It appears that during the past ten or fifteen years there was a tendency for anesthesiologists to justify their long period of training by using multiple agents simultaneously administered by complicated methods How can any but the most skilled control the course of anesthesia when a patient is receiving pentothal by vein, is breathing a mixture of several volatile agents and is paralyzed by a shot of curare? All the surgeon wants is simple unhurried light anesthesia in the majority of cases proper relaxation for abdominal work, minute-by minute control of pulse and respiration no cyanosis at any time, signs of waking up as the dressing is applied, and finally a rosy-colored, gently breathing patient wiggling a bit as he is removed from the operating room to the recovery room To obtain these objectives in each routine operation demands the utmost in devotion to the task at hand and vigilance undisturbed by activities in the operating room or distraction by visitors The surgeon is deeply dependent upon the anesthesiologist, who is responsible for carrying the child safely through

should be given are matters of continuing dispute For many years we have used scopolamine alone for infants, and morphine properly balanced with scopolamine for children above two months undergoing cardiac surgery and for children above six months undergoing routine surgery (see Chart) Scopolamine has completely displaced atropine as the proper drug to be used with morphine Dr William O McQuiston, our chief anesthesiologist, favors slightly higher preoperative doses of morphine, not only for routine cases, but also for all cardiac and thoracic cases, than those used in most children's hospitals Naturally, some judgment must be added to the directions given in the dosage chart, the skinny, vigorous youngster five years old will tolerate a larger dose than the fat lethargic child of the same age or the patient debilitated by prolonged disease Children properly medicated come to the operating room sleepy, mellow and cooperative, require less anesthetic agent and usually have little or no recollection of having gone to the operating room The dosages of morphine as outlined and used in many thousands of cases during the past twelve years have led to no case of alarming

PREMEDICATION FOR ALL SURGICAL CASES EXCEPT HEART, TONSILLECTOMY, ADENOIDECTOMY AND NEUROSURGERY

	Morphine Sulfate		*Scopolamine*	
Age	Mg	Grains	Mg	Grains
14 days			0 12–0 15	1/400–1/500
2 months			0 15	1/400
6 months	0 8	1/80	0 15	1/400
1 year	1 2	1/48	0 15	1/400
2 years	2 0	1/32	0 15	1/400
3 years	3 0	1/20	0 15	1/400
4 years	4 2	1/16	0 15	1/400
5 years to puberty	5 0–8 0	1/12–1/8	0 2–0 3	1/300–1/200
Past puberty	8 0–10 0	1/8–1/6	0 3–0 4	1/150

PREMEDICATION FOR HEART SURGERY AND OTHER INTRATHORACIC PROCEDURES

	Morphine Sulfate		*Scopolamine*	
Age	Mg	Grains	Mg	Grains
14 days			0 12–0 15	1/400–1/500
2 months	0 8	1/80	0 15	1/400
6 months	1 0	1/60	0 15	1/400
1 year	2 0	1/32	0 15	1/400
2 years	2 5	1/24	0 15	1/400
3 years	4 0	1/16	0 15	1/400
4 years	5 0	1/12	0 2	1/300
5 years to puberty	8 0	1/8	0 3	1/200
Past puberty	10 0	1/6	0 4	1/150

and died of suffocation Witnessing such a catastrophe leaves an indelible impression upon one s mind.

Local anesthesia, except for minor procedures such as sewing up small lacerations or removal of small superficial lesions has little place in children s surgery The apprehensive child screams when he sees the hated needle and rarely cooperates by lying still during the operation *Only in older children can it be used occasionally with some success* Local anesthesia produces undue strain on the child and plaques on the surgeon s coronaries

Curare likewise has little place in children s surgery It is Indian arrow poison which paralyzes all muscles and narrows the margin of safety during anesthesia. Somebody rightly said "Curare can make any anesthesia look good" What does not show is the danger of inadequate respiratory exchange and its fearsome consequence The use of curare in children except in unusual circumstances is condemned

Recapitulation

To decrease the blood pressure of stray anesthesiologists who may read this chapter it is appropriate to add that some of the foregoing statements are a bit too emphatic and not applicable under special con ditions and in some countries such as Britain, where even the smell of ether is offensive The hotly discussed subject of the role of the trained nurse-technician in anesthesia is appropriately bypassed except to say that many nurse anesthetists after years of experience have become uncannily capable Nevertheless the majority of nurses in pediatric anes thesia should, with few exceptions limit their anesthetic armamen tarium to the safest agent, ether and to the simplest methods of its administration. No objection is raised to the use of any agent or agents by the well trained physician anesthesiologist who knows the physiology of respiration, studies the pharmacology of drugs recognizes the need of constant, adequate exchange of gases and adheres to the virtues of sim plicity

A wise surgeon is not dictatorial about what anesthetic agent shall be used or by what method it shall be administered Surgeon and anes thesiologist ideally work in harmony are dependent upon each other and recognize and respect each other s skills The person at the head of the table must know from moment to moment the exact condition of the patient and be unafraid to advise whether operation can progress at the regular pace must be hurried or possibly discontinued. And the surgeon must make it possible for the anesthesiologist to bring the patient through the operation by wisely planning its extent and by observing the basic laws of minimal trauma and hemostasis Surgical stunts are as inappropriate in the operating room as anesthetic tricks Looking out

each operation by the simplest and safest measures known The spectacular has no place in anesthesia—just bring them back alive

For all routine operations in children we use open-mask ether. Ether has an odor repulsive to children, it is slow-acting, is slowly eliminated, produces postoperative nausea, and is old-fashioned, but it also has a characteristic which far outweighs all these objections it has a wide margin of safety In the hands of the average anesthesiologist and the trained nurse-technician, ether is undoubtedly the safest anesthetic agent known for children Until an anesthetic agent is found which will combine the safety of ether, the potency and speed of action of cyclopropane, the nonexplosive and odorless advantages of nitrous oxide, we shall continue to use good old ethyl ether for routine cases Cyclopropane anesthesia is used for all cardiac and thoracic operations Cardiac irregularities supposedly caused by cyclopropane do not appear in children

The fact that ether is given by open mask does not mean that the anesthetists close their eyes to all adjuncts of anesthesia Intubation is used for all open chest surgery and for most operations about the mouth, neck and throat, with the exception of tonsils A record showing two deaths—one unavoidable—in 29,000 consecutive tonsillectomies performed under ether and without intubation is sufficiently convincing that no change is warranted in anesthesia for tonsillectomy During every anesthesia a laryngoscope, tracheal tubes, suction, and so on, are always available for instant use in case the airway becomes obstructed

Some Technics We Don't Like

Spinal anesthesia has no place in children's surgery

We do not favor intravenous barbiturates except by those skilled in their use It is tempting to give an intravenous injection of pentothal for a short emergency procedure, such as straightening a green-stick fracture Many anesthetists routinely use this quick-acting agent in adults for induction of anesthesia and for short operative procedures We stubbornly refuse to use it in children because of the difficulty in accurately controlling the level of anesthesia and because of the danger of laryngospasm The argument that barbiturates may be given intravenously for short emergency anesthesia to a child who has just eaten is fallacious, because no child should be given any general anesthesia of any kind unless the stomach is empty It is a nuisance, but an essential one, to go through the unpleasant procedure of emptying the stomach before anesthesia is given A teen-ager with a fracture of both bones of the forearm was admitted to the emergency room of a hospital right after she had eaten lunch A hurried shot of barbiturate was given intravenously and the fracture reduced The child vomited, aspirated vomitus

and died of suffocation Witnessing such a catastrophe leaves an indelible impression upon one s mind

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of the window and absent-minded relaxation are appropriate only when the child is safely in the recovery room

The anesthesiologist with an audience of a half-dozen observers who has to spend more than a half hour in putting an infant to sleep because of unavoidable difficulties, and who during this time makes no excuses for his slowness and resorts to no drastic expedients to impress the onlookers or to console the impatient surgeon, is a gift beyond price to the welfare of children who are intrusted to his care in the operating room

Cardiac Arrest

Cardiac arrest is the most tragic complication that may arise during the course of an operation. Cardiac arrest in this discussion refers to sudden cessation of an effective heart beat during a major or minor operation in a child whose heart is basically sound.

The function of the heart is to beat, and so long as the heart is properly nourished by a continuous and adequate flow of oxygenated blood, and is not poisoned by excessive anesthetic agents carbon dioxide or drugs it will keep right on beating through any reasonable operation and thereafter with good fortune for a hundred years The heart is a tough piece of meat, only disease, starvation or poisoning will stop it.

Too much has been written about the treatment of cardiac arrest and too little about its prevention. Although no figures are available as proof I doubt whether anyone will disagree with this statement 95 per cent of cases of cardiac arrest occurring during operation on children—exclusive of heart operations—are preventable The other 5 per

cent would be preventable if surgeons and anesthetists were not erring human beings

Prevention

Prevention of cardiac arrest is aided by attention to a number of details

FIRST, AN EMPTY STOMACH. To enforce the dictum that a general anesthetic will not be given to a child unless the stomach is empty will require constant vigilance and cooperation by the entire hospital staff A child in a ward will beg food or water from the other children the morning of operation Before anesthesia is begun on any child with obstruction between the pylorus and the rectum the stomach must be aspirated with a proper-sized catheter even though the patient is a newborn infant who has never received fluid or food For patients with intestinal obstruction it is wise to keep a catheter in the stomach during the entire operation Manipulation of distended bowels may cause reflux into the stomach and esophagus and aspiration into the tracheobronchial tree A tiny piece of food or clotted milk aspirated during anesthesia may easily cause respiratory obstruction and fatal suffocation Signing the death certificate with the high-sounding term "cardiac arrest" does not assuage the remorse for not having taken the precaution of emptying the stomach before anesthesia was begun

SECOND, A FREE AIRWAY Whether inhalation anesthesia is being given by open mask or by intubation, easy unobstructed breathing is essential Children with chronic productive bronchiectasis, infants with tracheo-esophageal fistula complicated by bronchopneumonia, and those few children who constantly have excessive mucus in the tracheo-bronchial tree will have to have aspiration frequently during the operation to keep the airway open The slightest sign of cyanosis is an indication for immediate investigation and correction To the anesthetist we say, "Never mind the itchy surgeon who makes impatient comments Stop the anesthesia, give oxygen and re-establish normal color at once" A mother flies into a panic if her child gets blue, the child, unaccustomed to cyanosis, gasps for air Just because the child under anesthesia does not raise a fuss about oxygen want is no evidence that his physical mechanism is not suffering It is too bad that the cyanotic anesthetized patient cannot scream—many cases of cardiac arrest would be avoided

THIRD, AVOIDANCE OF TOO-DEEP ANESTHESIA. All anesthetic agents are poisons Professor Tatum in his lectures on pharmacology at the University of Chicago used to repeat with solemn emphasis, "Remember, when you anesthetize a patient you half kill him" How much anesthetic agent is necessary for any given operation and how fast it may be administered are matters of experience and judgment on the part of the person

at the head of the table The surgeon can cooperate by refraining from critical remarks or showing signs of impatience if at times the anes thesia is too light

Abdominal surgery in children can be performed under relatively light anesthesia except during that short time while the peritoneum is being closed. All other operations including intrathoracic procedures are best done under continuously light anesthesia. For heart operation on very blue infants with tetralogy of Fallot the anesthesia should be so light that when one s finger is placed in the palm of the patient s hand, the fingers will close Some anesthetists are apologetic if a patient wiggles a bit from time to time Wiggling may be injurious to the ego of the anesthetist, but is far safer than cadaveric relaxation

FOURTH, REPLACEMENT OF BLOOD LOSS Responsibility for main taining adequate blood levels during every operation is shared by the surgeon and the anesthetist. The surgeon must be constantly aware of blood loss and must from time to time check on the number of sponges used and the amount of blood in the suction bottle Weighing bloody sponges is a bit of a nuisance and not too accurate but it is well to go through the procedure occasionally to refresh one s mind on the quantity of blood a standard sponge will absorb Blood loss is usually underesti mated. In these days of easy availability of blood there is little excuse for allowing a patient to go into shock. Blood loss results in hypoxia, if the loss is great and is not replaced, the most susceptible tissue—heart muscle—stops because of lack of fuel. Once again, entry of the high sounding term "cardiac arrest" on the death certificate does not alter the fact the death was due to just plain hemorrhage

Measurement of Pulse, Blood Pressure and Respiration

A good pulse and a steady blood pressure are excellent indicators of adequate circulation during adult anesthesia, but in infants are less reliable. Often it is impossible to get any measurement of blood pressure in an infant, and evaluation of the pulse is inaccurate except by the most experienced Recently machines have been devised for monitoring the heart beat during operation. A gadget is applied to the patient s chest, and the anesthetist hears the machine click or watches a needle swing back and forth with each heart beat. Unfortunately the only indication of impending cardiac arrest shown by such a machine is slowing of the pulse

It is equally important to know from moment to moment whether air is going into the lungs Both pulse and respiration are best monitored by taping a stethoscope over the site of the apex of the heart. Another advantage of this method is that it keeps the anesthetist at the patient s head all the time.

It is an accepted fact, uncolored by a desire on the part of the surgeon to escape responsibility, that the anesthetist (technician) or anesthesiologist (physician) is responsible for practically all cases of cardiac arrest in all operations except those on the heart itself In spite of thorough understanding of methods of prevention, cardiac arrest will occasionally occur—often when least expected It should be rare During the past twelve years we have had six unanticipated cases of cardiac arrest as defined in the first paragraph of this chapter Excessive anesthesia and inadequate respiratory exchange were to blame for all Five of the six cases occurred in infants Cardiac massage through the opened chest and intracardiac drugs such as calcium lactate and epinephrine were used, but to no avail Four children died and, unfortunately, two survived, unfortunately is the proper descriptive word because these two children are unconscious metabolizing masses of flesh To see a child who because of cardiac arrest now lies spastic and unseeing in its crib, and to witness the anguish of parents slowly realizing that their child will never again be normal, will drive those persons who officiate at the head of the operating table humbly to vow that such a tragedy shall not recur

Postoperative Care

As the last skin stitch is tied, postoperative care begins At this very moment, after a long and tedious operation, vigilance is apt to relax. The tired surgeon pulls off his gloves and walks out of the room, and the resident places a dressing over the wound and quickly follows the surgeon to the staff room for a cup of coffee leaving scrub nurse and anesthetist to move the patient to bed or crib and transport him to the recovery room. During this brief period many things may happen A drainage tube may be pulled out of the chest as the child is lifted from the operating table· the polyethylene tube may inadvertently be jerked out of the saphenous vein the bottle of blood reached for by a too short nurse standing on tiptoe, crashes to the floor the child suddenly vomits, and the anesthetist hasn t enough hands to turn the patient, lower the head of the table and aspirate the pharynx all at once the scrub nurse becomes enraged when the resident nonchalantly walks out of the operating room and leaves to her and the anesthetist the task of moving a heavy patient from the operating table to the bed All these catastrophes—mostly minor

—will occur from time to time unless the surgeon or the surgical resident stands by until the child is safely on his way to the recovery room, properly attended.

The Recovery Room

This room, advantageously on the same floor as the operating room to avoid delays at elevators, serves the most important function in postoperative care Fortunate is the surgical staff blessed with well trained, resourceful nurses in charge of this service

Infants and children are transported in crib or bed to the operating room—never on carts or stretchers—and are returned to the recovery room in the same crib or bed Routinely, the child is placed on its side when leaving the operating room and kept in that position until conscious Infants are supported in this position with sand bags so that vomitus and nasopharyngeal secretions may run out of the side of their mouths Restraints are necessary for children receiving intravenous fluid, but no child is ever completely restrained in the supine position It is not difficult to imagine what might happen to a semi-conscious infant who suddenly vomits in such a restrained position

Wall suction and oxygen are at the head of each bed Oxygen is usually given after most major operations For routine cases a funnel discharging a stream of oxygen is placed in front of the child's nose and mouth, infants are placed in Isolettes and lie in an atmosphere of 30 to 40 per cent oxygen, older children who have had major surgery are placed in oxygen tents Suction is used freely to keep the pharynx clear of mucus, blood and vomitus

A large portable cabinet containing every item necessary for respiratory emergencies is always at hand On the top of this cabinet, in a Craftsman's tool chest purchased from Sears, Roebuck and Co, are kept laryngoscopes, infant and child size, intratracheal tubes, various-sized plastic suction tubes, catheters, thumb control aspirating tips, airways and a face-fitting anesthetic mask for giving oxygen under pressure A gauge for controlling oxygen pressure is attached to the side of the cabinet. In the drawers are kept extra tubes, catheters, suction tips, 50-cc syringes and needles, sterile dressings, an emergency set of a few essential instruments, sterile gloves and all sorts of emergency drugs. In the lower drawer are complete tracheotomy sets, infant- and child-size

Routine cases, such as herniotomies, appendectomies, circumcisions, and the like, require close observation only until consciousness has returned In an hour or less these patients are returned to their rooms or wards anywhere in the hospital as conditions and parental wishes de-



mand More difficult cases, such as thoracotomies, atresias of the gastrointestinal tract, bowel resections, and so on, are kept in the recovery room for longer periods—sometimes all day

The child who has had an operation for congenital heart disease invariably has a drainage tube in his chest connected with a water-seal bottle The bottle is set on the floor under the bed and fixed with strips of adhesive tape to avoid overturning The moment the connections are made, the water level in the bottle is marked on a piece of adhesive stuck to the side of the bottle and the exact time written next to the mark With a ruler as a guide pencil marks are made on the adhesive tape above the water line at ¼-, ½- and 1-inch intervals Identical-size bottles, in which a rise in the fluid level of ½ inch equals 100 cc, are used so that the observer may know at a glance the exact amount of drainage The fluid level is frequently checked, and any rise of ¼ inch or more is immediately reported and investigated One expects drainage of 25 to 50 cc after any thoracotomy If bloody drainage exceeds reasonable amounts, it is replaced by transfusion, cubic centimeter for cubic centimeter A continuous rise eventually calls for reopening the chest and searching for a bleeder Fall in blood pressure and weakening pulse without a rise in the drainage bottle call for examination of the tube, chest examination and occasionally a roentgen film to be sure that blood is not accumulating in the chest

The pulse and blood pressure are checked in older children every fifteen minutes for a few hours, or longer if necessary Color and respiration are watched with hawklike attention Infants require little postoperative sedation, older children receive codeine or relatively small doses of morphine as indicated The electrode of a telethermometer is left inserted in the rectum for easy observation of the temperature A rise or fall in temperature is controlled by running warm or cold water through a Bigler rubber water mattress placed beneath the patient

The second recovery room is a phrase coined to describe an adjoining area of two- and four-bed wards on the same floor accommodating twenty patients The seriously ill postoperative patients, including all heart cases, are moved from the recovery room to these wards as their condition allows Regardless of color, race or financial status, all infants and children requiring close observation are moved to these wards, where another group of trained nurses is in constant attendance Occasionally, wealthy parents (rare in pediatric surgical practice) request, but are refused, special nursing care for their child in a private room after operation When it is explained that the child will be safer in these specially supervised wards, all objections vanish Patients are moved from this second recovery room area to other floors as demand for beds arises

Suffocation

Suffocation due to aspiration of foreign material into the tracheobronchial tree is the most common cause of postoperative death in all types of pediatric surgery. Infants and debilitated little children are in danger not only during the immediate postoperative period but also when feedings are begun. In fact, all infants weakened by disease, whether operated upon or not, are liable to vomit, aspirate and choke to death. For this reason special equipment is always available and teaching is directed toward its proper use. All surgical residents must familiarize themselves with the use of laryngoscopes and learn the trick of quickly inserting an intratracheal tube. As a matter of fact, it would be well for every resident in pediatrics and all its subdivisions to become expert with the laryngoscope. Because aspiration may occur at any time an emergency kit is kept on every floor. In this kit are an infant laryngoscope, intratracheal tubes, 2 catheters and 2 plastic tubes for suction, a 50-cc. syringe and needles and a face fitting mask for administration of pressure oxygen. The box is about the size of a fishing box purchasable at any hardware store. It is the duty of the head nurse to check the equipment daily and to be sure that all student nurses on the floor know where it is kept. Loss of equipment by pilferage or just plain vanishing into thin air represents a problem to the administration. Thus far it has been possible to convince the purchasing office that keeping the emergency kit in a locked cabinet defeats the purpose for which it is intended. Besides the impressive-looking stethoscope hanging from his neck, every resident in a pediatric hospital might well carry a laryngoscope in his back pocket.

Tracheobronchitis

Tracheobronchitis is a troublesome, although usually not serious, postoperative complication. As soon as a child begins to cough he is placed in an atmosphere of high humidity in an Isolette or oxygen tent. If the cough is dry and persistent, the patient is moved to the "steam room" in saturation humidity. Actually the air in this room is saturated with vaporized cold water but the room retains its name from the days of steam. It is gratifying to see how rapidly a cough which will not be quieted by generous doses of codeine will subside in 100 per cent humidity.

Patients who have been operated upon for atresia of the esophagus with tracheo-esophageal fistula, anomalies of the aortic arch and closure of interventricular septal defects are most apt to have postoperative tracheitis and edema leading to laryngeal or high tracheal obstruction. It is a matter of fine judgment to decide whether or when such a child

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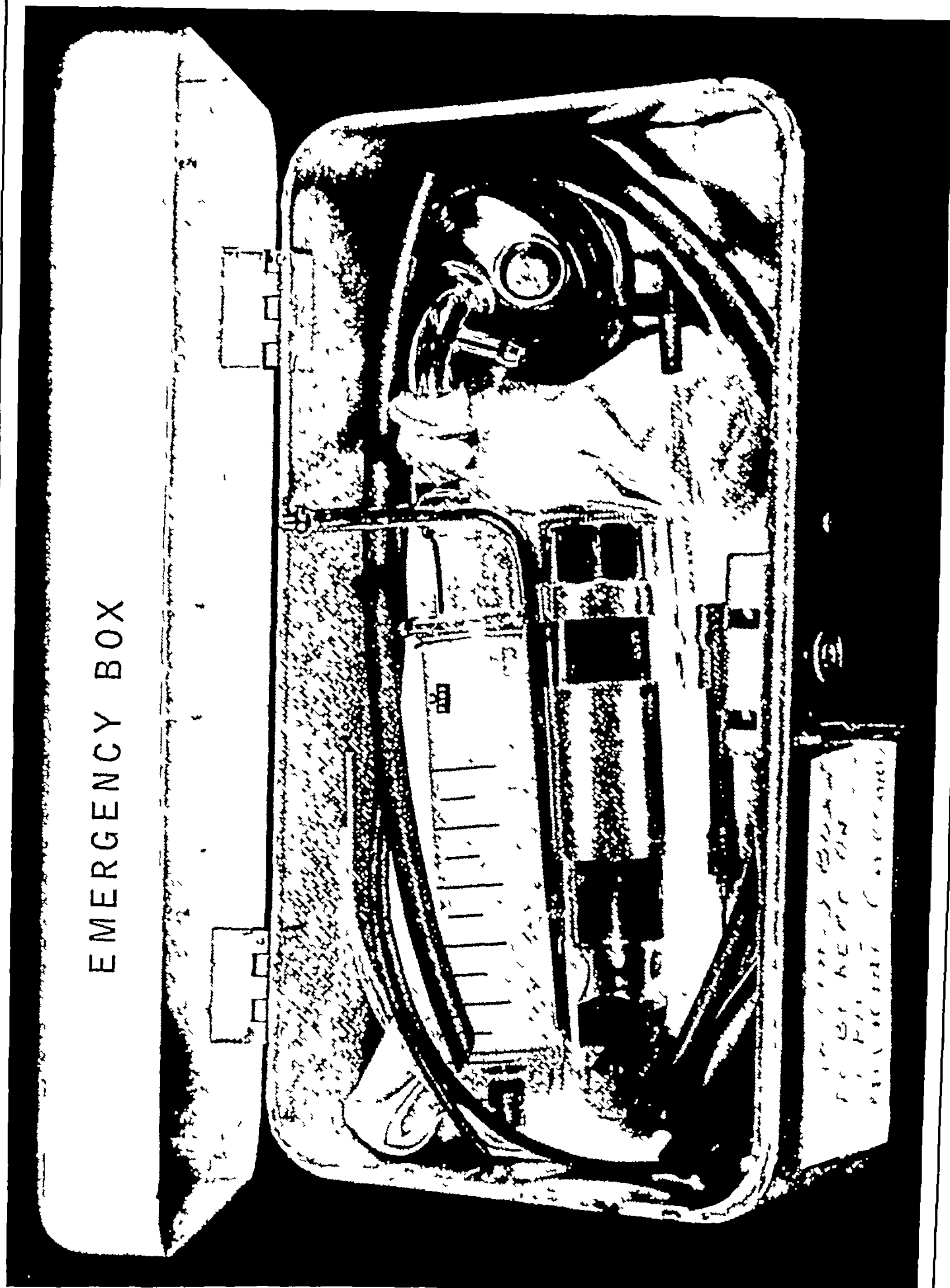
requires a tracheotomy When labored breathing with suprasternal retractions appears and persists, and the child shows signs of exhaustion it is high time to do a tracheotomy Although tracheotomy especially in an infant, is in itself a serious procedure, we have in the past erred in putting it off too long. The child struggling for air is watched continuously by resident and/or attending surgeon the operating room is readied, and, in the absence of improvement but well before complete exhaustion has occurred, a tracheotomy is done Each time one sees the sudden change in a child now falling asleep and breathing easily through the tube one wonders why the tracheotomy was delayed as long as it was

Postoperative Atelectasis

Postoperative atelectasis commonly follows not only thoracotomy but also any operation of magnitude especially in the very young and debilitated Again aspiration of mucus, thick saliva and particles of food is the primary cause Children will not cough voluntarily after a chest operation If excessive mucus accumulates in the throat or if the breathing is raucous the throat is aspirated frequently with a catheter Touching the back of the pharynx will stimulate some gagging, but will also often start involuntary coughing. Atelectasis demonstrated by physical examination and confirmed by roentgenogram is not a serious problem in children Usually in the course of a few days the lung expands spontaneously if not, the endoscopist is asked to insert a bronchoscope and clear out the obstruction Blowing up balloons or blowing water from one bottle to another is fun for the children and helps expand the lung.

Antibiotics

Antibiotics are used excessively in most hospitals after operation To prevent postoperative infection from arising unexpectedly the surgeon is apt to be too liberal in the use of antibiotics for patients who have had "clean" operations Although antibiotics are not given to patients who have simple operations, such as herniotomy excision of thyroglossal duct cysts, pyloromyotomy simple appendectomy and so on, we weakly conform to custom and give them to children who have had what is now a relatively common operation—division and suture of a patent ductus In fact, all children undergoing heart surgery receive 300 000 units of penicillin twice daily beginning the evening before operation and continuing for approximately 5 days after operation In patients who have infected wounds due to unavoidable gross contamination or to misfortune we use antibiotics freely Until the organisms have been isolated and sensitivity tests run we give "shotgun" doses of antibiotics hoping that one will hit The efficacy of these phenomenal drugs is


EMERGENCY BOX

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miraculous, but probably during the next ten or twenty years we will regret their too liberal use. Already the Staphylococcus is beginning to show signs of defiance. In fact, staphylococcal infections in infants which ten years ago responded promptly to penicillin now thrive on it.

Gastric Suction

Gastric suction after abdominal operations of considerable magnitude, especially for partial or complete intestinal obstruction, is important for the re-establishment of bowel tone and for the prevention of aspiration of vomitus. The ordinary Levin rubber tube, size no. 8 or 10 French, inserted through a nostril into the stomach is satisfactory for suction for a day or two, but should not be left in longer because rubber is irritating to the nasal mucosa and may cause necrosis. Plastic disposable tubing is more satisfactory and may be left in for a number of days. However, change to the opposite nostril is still wise if prolonged suction is necessary. Negative suction on the tube should be cut down considerably below that ordinarily delivered by a suction machine. We have had two cases of perforation of the stomach during prolonged gastric suction. Whether the mucosa was caught in the end of the tube and underwent necrosis or whether the tube was pushed through the stomach wall we were unable to determine at postmortem examination. It could be that the end of the tube pressing against the stomach wall caused necrosis and rupture.

It is well to give the infant or child who has a tube in the stomach a teaspoonful of water fairly frequently, not only to maintain patency as the water returns, but also to keep the mouth clean. Incidentally, children are grateful for the pleasure of a sip of water from time to time.

When to remove the tube and start feeding is a bit difficult to decide in infants. If little or no material returns through the tube for approximately six hours, it is well to remove the tube and start sips of glucose water. Allowing an infant to have more than sips of fluid while the tube is still in the stomach, but clamped off, is not satisfactory because irritation of the throat by the tube is apt to cause gagging and vomiting. One thing is sure: so long as bile-stained material is being extracted from the stomach, gastric suction must be continued.

Feeding

There is a tendency among residents to withhold food too long. The high metabolism of an infant demands that feedings not be withheld any longer than absolutely necessary. As soon as the baby looks alert and sucks his fist he is usually ready for cautious feeding. When he

responds to attention with a smile—provided he is six weeks old or more—a regular formula may be ordered without hesitancy The pacifier frowned upon for many years as an indulgence is coming back to its rightful position as a source of enjoyment for the infant and, I believe as a therapeutic measure Every infant loves to suck on anything, and a good healthy workout on a pacifier appears to be a stimulus to appetite. The notion that a baby fills his stomach with air when sucking on a pacifier has been refuted. The contentment gained from a pacifier is certainly better than the exhaustion caused by prolonged crying Let the little "guy" have some fun.

Wound Dehiscence

This should be less frequent in children than in adults because of lesser strain on the wound and the uninhibited healing power of young tissue, but it is not Muscle splitting incisions used in the right upper quadrant for pyloric stenosis and in the right lower quadrant for appendectomy and intussusception practically never rupture, but long trans rectus or paramedian incisions especially if infected, occasionally let go Change from running catgut to interrupted silk sutures for closure of the fascia has lessened slightly the danger of wound dehiscence

Vomiting, distention, sudden pallor or shock following abdominal surgery calls for examination of the wound. If the dressing is saturated with bloody serum, even though one sees no loop of bowel, the wound has given way beneath the skin. It is well under such circumstances while removing the skin sutures, to have ready a large sterile dressing poised above the wound to catch the bowels before the crying child forces them all over the abdomen and onto the bed The dressing is fixed with adhesive tape and preparations are made for immediate closure. Closure of disrupted wounds in children is most successfully accomplished by a single row of sutures including all layers of the abdominal wall We use double fine wire sutures attached to small buttons The double suture with a button attached at one end is carried through the skin muscle and the edge of the peritoneum and brought out in a similar manner on the other side of the wound. A second button is threaded onto the sutures After all the sutures have been placed they are drawn up snugly enough to appose the raw edges and tied. Second rupture of the wound rarely occurs The sutures are left in for ten to eleven days If the ruptured wound is grossly infected, there will be some tendency for the buttons to produce small areas of skin necrosis This complication is unavoidable but far less troublesome and disfiguring than the transverse pressure necrosis cuts caused by through and through sutures of silk or wire without buttons

Dressings

It has been difficult to break the long-established habit of piling a mass of dressings on all operative wounds A single folded sponge held in place by a few adhesive strips is sufficient Massive bandages serve no useful purpose and are a hindrance to satisfactory abdominal or chest examination Large dressings over lower abdominal wounds in infants are soon saturated with urine For all children below teen age it is well to seal the dressing with adhesive tape on all four sides to keep curious fingers from exploring one's handiwork

Suture Removal

Removal of skin sutures, usually about the seventh day after operation, is a minor procedure for the surgeon, but a trial to the child For a child old enough to follow simple conversation it is explained that the dressing is going to be changed and that a few tiny stitches are going to be removed Not fully grasping what is to occur, the child logically asks, "Will it hurt?" A trick, often effective, is to answer that question by catching a hair on his head, giving it a little pull, and telling him it will hurt about that much "If you lie real still it will hurt less" This suggestion is worth trying, but not always effective The scissors must be of the embroidery type with very fine points so that the sutures can be cut without pulling Finally, after a bit more distracting conversation the end of one of the sutures is slyly cut off and shown to the child A little time, plenty of reassurance and the utmost gentleness to avoid pulling will usually allow suture removal without crying and struggling The child is relieved when the ordeal is over and so very proud that he didn't cry His ego is inflated when the surgeon says, "You sure are a good boy"

Some children are impossible, scream madly the moment the dressing is touched, and fight like demons during the entire procedure In such instances there is nothing to do but have a few people hold the child while the stitches are being removed Occasionally, when a sudden jerk during removal of sutures from a skin graft, for example, might dislodge the graft, it will be necessary to quiet the child with general anesthesia

Thyroglossal Duct Cyst

Diagnosis

A persistent rounded mass in the midline of the neck over the region of the hyoid bone is probably a thyroglossal duct cyst. Such cysts are usually seen in children between the ages of one and five years but may be present at birth or may not appear until adulthood. In typical cases the child is brought in because of the symptomless lump A mass not tender usually about 1 cm in diameter rather firm, sometimes cyst like is felt over the hyoid bone and can be seen to move up and down as the child swallows or sticks out his tongue

A condition mimicking a thyroglossal duct cyst is a midline submental lymph node Fixation of the gland to the subcuticular tissue over the hyoid bone as a result of previous chronic inflammation makes an immediate differential diagnosis impossible A sebaceous cyst in this area is easily recognizable by its position in the skin and its mobility There should be no difficulty in distinguishing an uncomplicated thyro-

glossal duct cyst from a branchial cyst, because the former always lies in the midline, the latter never A thyroglossal duct cyst which extends slightly towards one side or the other from the midline does so because it has been drawn over by scar tissue, the result of previous inflammation, or because the expanding cyst has bulged towards one side. At operation one will always find the base of a thyroglossal duct cyst attached to the mid-portion of the hyoid bone

A persistent sinus over the middle of the hyoid bone continuously discharging some mucus, together with a history of a local abscess which drained spontaneously or was opened by a physician, makes a diagnosis of thyroglossal duct sinus or fistula unmistakable

A few unusual variations of thyroglossal duct cysts occur A cyst may be present at the base of the tongue in a newborn infant and cause respiratory distress Coarse, noisy, gurgling breathing, suggesting pharyngeal obstruction, calls for palpation of the base of the tongue

In one instance a golf ball-sized cyst in the neck of a twenty-four-hour-old infant required emergency operation to relieve acute respiratory distress caused by pressure against the anterior wall of the pharynx In rare instances the cyst, instead of overlying the hyoid bone, lies over the thyroid cartilage or even in the suprasternal area

Treatment

Treatment of thyroglossal duct cysts and sinuses is surgical Removal of the cyst or sinus without excision of the midportion of the hyoid bone will surely result in recurrence

Under general anesthesia, with the child's head well extended, a 1½-inch transverse skin incision is made over the cyst, or an oval incision is made around the sinus After suitable exposure a bit of methylene blue is injected into the intact cyst Demonstration by the anesthetist of blueness at the base of the tongue is an impressive teaching procedure and incidentally helps to identify the fistula in the muscles of the tongue between the hyoid bone and the foramen caecum In the majority of cases, however, methylene blue will not go beyond the hyoid bone, and this fact must not be interpreted as evidence that superficial excision of the cyst will be curative It is usually impossible to inject methylene blue into an open sinus

The cyst is then grasped with an Allis forceps and held up under tension to facilitate separation of muscle from the mid-portion of the hyoid bone With a small curved hemostat the posterior surface of the hyoid bone is separated from the structures beneath Danger of breaking into the pharynx is minimal if the dissection is kept near the under surface of the bone With a stout scissors or a curved bone-cutting forceps a 1-cm wide section of the middle of the hyoid bone is excised The

cyst and attached segment of bone are gently held up to avoid breaking and losing the tract as it is carefully followed through the myelohyoid muscle to the foramen caecum.

At this point the anesthetist is asked to push down with her finger on the back of the tongue so that the fistula may be followed to its origin and ligated at that point The operation has been completed. Actually it is unnecessary to bring together the ends of the hyoid bone but we prefer to do so to avoid accumulation of a blood clot in the open space The muscles and subcuticular tissues are snugly apposed with interrupted fine silk sutures and the skin is closed with a fine nylon suture The wound is never drained The skin sutures are removed on the third rather than the seventh day after operation to lessen scarring of the needle puncture.

The technique of removal of a thyroglossal duct sinus or fistula is the same as outlined above

At times when it is difficult to determine whether the exposed mass in the neck is a cyst or a broken-down lymph node, the string test is applied. The mass is opened, and the thick liquid content is touched or grasped with a forceps If a sticky string of mucoid material adheres to the instrument as it is withdrawn, a diagnosis of thyroglossal duct cyst is established

It is well to examine carefully a mass which upon exposure just below the hyoid bone appears to be solid tissue It may be thyroid, and what is more important, it may be the only thyroid tissue the child has Such a nodule should be split in half in situ and, if not thyroid tissue grossly a frozen section should be examined. If the mass proves to be thyroid, each half of the gland with its blood supply intact should be tucked beneath the muscle on each side of the neck.

The acutely infected thyroglossal duct cyst is treated as any other infection. Usually an abscess forms which has to be drained or opens spontaneously Sometimes after a week or two of drainage, the wound closes spontaneously Operative excision is done at a later date after all signs of infection have disappeared and the cyst is beginning to re-form More often a mucus-discharging sinus persists after the infected cyst has been drained. After a month or two when it is obvious that the sinus will not close, operation is advised.

The rare thyroglossal duct cyst at the base of the tongue may usually be cured by needle aspiration alone If the mass at the base of the tongue is solid tissue a biopsy should be done rather than an excision. In one case a mass in this location was removed it proved to be normal thyroid tissue, and soon thereafter the patient showed signs of myxedema.

Recurrence of thyroglossal duct cysts after operation is common and has only one cause failure to remove the middle of the hyoid bone

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Embryologically, a thyroglossal duct cyst is a remnant of the thyroid stalk extending from the foramen caecum to the thyroid. This stalk always goes anterior or posterior to, or through the middle of, the hyoid bone. Can there therefore be any excuse for not removing the involved section of this small bone?

Congenital Atresia of the Esophagus

Congenital atresia of the esophagus with tracheo-esophageal fistula is one of the most difficult deformities the pediatric surgeon is called upon to treat. Delayed diagnosis, pulmonary complications, other congenital defects underweight and prematurity all contrive to keep the surgeon humbled by a surgical mortality rate of 30 to 50 per cent

Diagnosis

The diagnosis is easily made if the obstetrician thinks of the possibility in a newborn child who immediately after birth chokes, coughs and has an excessive amount of secretion in the nose and throat. However one shouldn t criticize the obstetrician for missing a diagnosis since

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Pressure on the anesthetic bag will have to be gentle or the stomach will be enormously filled with air

A plastic tube is inserted into the saphenous vein at the ankle for administration of fluid and blood The infant is placed in the prone position with the right chest slightly elevated Through a posterolateral parascapular incision the chest is opened through the fourth intercostal space For years we attempted to enter the chest through a somewhat similar incision *extrapleurally* but gave up this approach because about half the time in spite of the utmost care, the pleura was accidentally opened Furthermore the transpleural approach reduces the operating time by about one-half hour and affords better exposure

The lower end of the esophagus is quickly identified, dissected free clamped at its connection with the trachea and cut. The lower end is left open to allow accumulated air in the stomach to escape To this point the anesthesia probably has not been too satisfactory After the tracheo-esophageal fistula has been clamped the anesthetist can properly expand the lungs and aspirate secretions from the trachea and bronchi If as is usually the case the right upper lobe of the lung is atelectatic, a bit of gentle massage may force occluding secretions into the bronchus from which they may be removed by aspiration

The stump of the esophagus on the trachea is closed with four or five interrupted stitches of 5-0 silk. Closure of the fistula too close to the trachea may later cause tracheal stenosis Leaving a long stump of esophagus on the trachea may later result in the formation of a pocket in which secretions collect

The upper end of the esophagus is thoroughly freed from surrounding structures With an Allis forceps on its lower tip it is pulled down as far as possible and thoroughly freed from surrounding structures A ductus clamp is applied across the upper end, and the tip slightly damaged by the Allis forceps is cut off The mucous membrane is opened and dissected from the muscularis for a few millimeters to allow better identification during anastomosis The lower end of the esophagus is grasped with another ductus clamp and the ends are brought together for suture (Be sure the clamps are completely closed to the third notch or the muscularis may tear)

Much difference of opinion exists about the best method of suture two-layer or one-layer anastomosis? many sutures or few? 5-0 or 6-0 silk? Our method of suture finally stabilized and routinely used for the past five years is not simple technically but is effective We believe we may say effective without danger of criticism because we have had only two leaks in the past four years in approximately fifty cases The mucosa is coapted with interrupted sutures of 6-0 Deknatel silk on a swaged-on needle The stitches usually eight to ten are so placed that the knots are inside the lumen of the esophagus We believe that better healing occurs

the deformity is rare—probably less than one in 2000 births. One would hardly expect a busy obstetrician who has delivered, for example, 1500 infants without esophageal deformity to pick off the diagnosis in the fifteen hundred and first infant. And yet, in the interest of early diagnosis, it is urged that, when suspicion of obstruction exists, the obstetrician attempt to pass a catheter. If it meets obstruction, the diagnosis is practically made. In the majority of cases the diagnosis is not suspected until the baby has been moved to the nursery and feedings are begun. Prompt regurgitation of all fluid and formula which is not sour or coagulated further calls attention to the probability of esophageal obstruction. It is not unusual—and is at times a bit disconcerting—to have a student nurse fresh from the classroom where diseases of the newborn have been discussed gently suggest the correct diagnosis.

The infant suspected of having esophageal obstruction is moved to the x-ray department, where an upright film is taken of the chest and abdomen in the anteroposterior and lateral positions to determine the condition of the lungs and to demonstrate the presence or absence of air in the gastrointestinal tract. One cubic centimeter of Lipiodol—warmed so it will flow—is injected by catheter into the upper end of the esophagus. The typical blind pouch at the level of the second, third or fourth thoracic vertebra clinches the diagnosis. By no means should barium be used for this procedure; it may be aspirated into the lungs, where it will cause bronchial irritation. If the intestine contains air, a fistula exists between the lower segment of the esophagus and the trachea. Contrariwise, absence of air in the gastrointestinal tract suggests atresia of the esophagus without fistula. Incidentally, close attention to air patterns in the bowel will often aid in making a diagnosis of bowel atresia—a not uncommonly associated deformity.

After the diagnosis has been established preparations are made for operation. During these few hours the pharynx is aspirated approximately every fifteen minutes. Aspiration not only removes constantly accumulating saliva and mucus, but also stimulates cough which may free some secretions from the lungs. Suction must be gentle and is best done with a thumb-controlled aspirator attached to a catheter. Too vigorous aspiration will cause local edema and troublesome bleeding from the mucous membrane.

Operation

The infant is anesthetized with open-mask ether. Because the respiratory exchange is lessened by pressure of a stomach full of air against the diaphragm, anesthesia will be obtained slowly and the surgeon will have to be patient—a real trial. As soon as the baby has relaxed, an intratracheal tube is inserted and anesthesia continued with cyclopropane

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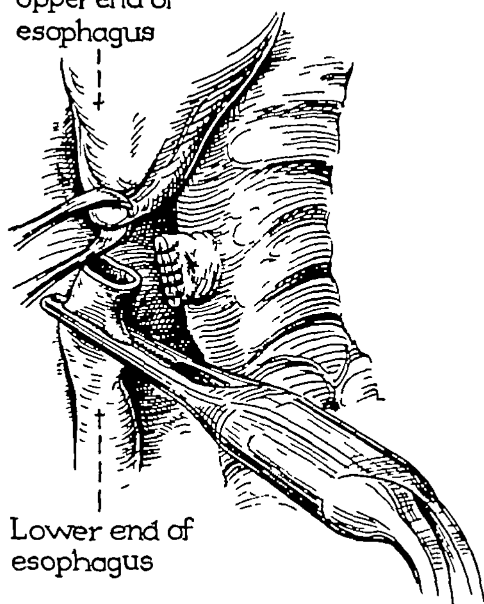
After the anastomosis has been completed a catheter with a number of extra holes near the end is introduced through a tiny stab wound through the sixth interspace as near the spine as possible The end of the catheter is brought near the anastomotic site in the esophagus and fixed there with a catgut suture The wound is closed in layers and the infant returned to its humidifed and oxygenated crib All these infants regardless of financial circumstances of their parents are cared for in one ward by specially trained nurses The drainage tube is connected with a water seal bottle

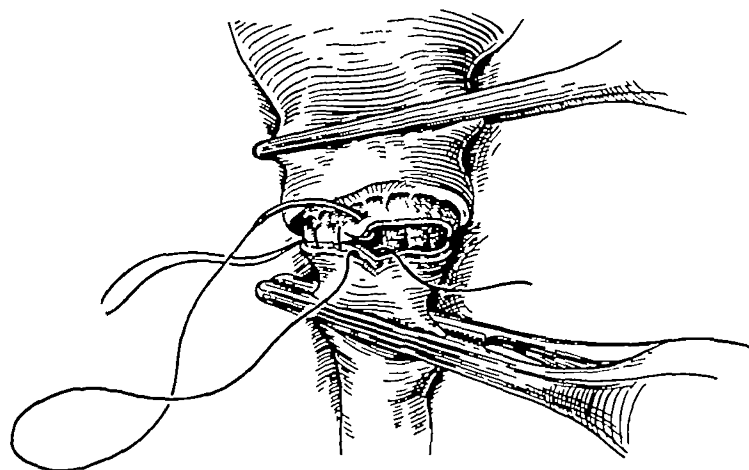
Postoperative Care

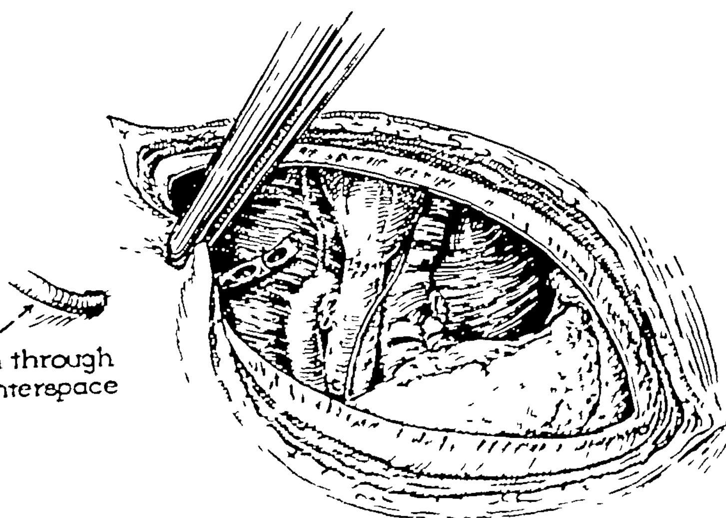
Postoperative aspiration is performed as indicated—every hour if necessary Every infant who has had a tracheo-esophageal communication even for a day or two has bronchitis due to the irritation of regurgitated gastric juice Painstaking and gentle removal of consequent obstructions is unquestionably the most important feature of early postoperative care The distance from the lips to a point a bit short of the site of anastomosis is measured and marked on a catheter with a few layers of adhesive tape The nurses are instructed when aspirating the pharynx never to insert the catheter beyond the adhesive marker If the child has much sticky material in its throat, it is helpful to stimulate the infant to cry by snapping the soles of the feet. Crying is often effective not only in releasing secretions, but also in helping to prevent or clear atelectasis A laryngoscope suction machine, intratracheal tubes catheters and aspirating tips are constantly at the baby's bedside If respiratory embarrassment or cyanosis develops a surgical resident familiar with intubation is given an emergency call Quick intubation and removal of small slugs of tenacious mucus have spared us many catastrophes

Nothing is given the baby by mouth until about the fifth day when a swallow of Lipiodol is observed roentgenographically If it readily goes into the stomach and no leak is demonstrated feeding is carefully begun

It seems strange that people—in this instance we the surgeons—can be so stupid so long. We used to feed these children their formula with bottle and nipple according to the standard fashion of infant feeding We did add to the order a note that the feeding should be given slowly That was not enough A hungry infant nursed vigorously and literally drowned in his formula. One can easily understand why The dilated upper end of the esophagus filled with milk faster than it could escape through the tiny lumen of the still undeveloped lower esophagus Sudden


Upper end of
esophagus
Lower end of
esophagus




Drain through
6th interspace

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regurgitation was followed by aspiration into the lungs and consequent respiratory obstruction. Eventually—but too eventually—we learned to feed these infants slowly with a medicine dropper. It is now routine for the nurse to hold the infant on her lap for feedings. The medicine glass containing the formula is placed on a stand. The nurse fills the medicine dropper and gives it to the child. The time it takes the nurse to reach over and again fill the medicine dropper is sufficient to allow the previous amount given to pass through the esophagus. As a further safeguard an aspirating machine with catheter and suction tip, all connected, is ready for use in case the baby regurgitates or coughs. This simple process of slow feeding has reduced postoperative pulmonary complications and lowered the mortality rate remarkably. Such feeding is continued until it is demonstrated that the baby is ready for bottle and nipple feeding.

Upon discharge from the hospital the parents are instructed to note and report any difficulty in swallowing and are asked to return at three months for barium studies of the esophagus. After uncomplicated healing of the repaired esophagus stricture rarely occurs and dilatations are seldom necessary.

Gastrostomy is performed *only* if a sizable leak occurs at the anastomotic site in the esophagus. The gastrostomy then is necessary for feeding and later is advantageous for dilatations of the almost inevitable stricture. Before the stricture has become all but occlusive preparations are made for dilatations. A long piece of no. 2 braided silk is tied to a fine catheter which is guided through the esophagus into the stomach under fluoroscopic control. The catheter is fished out of the stomach through the gastrostomy, and the thread is pulled through. The ends of the thread are tied together. Dilatations are performed in the following way. The thread is cut, and the end near the gastrostomy is tied to a bougie. To the other end of the bougie a new thread is tied. By grasping the end of the thread near the mouth the bougie is pulled in retrograde fashion through the stricture and out of the mouth. As the bougie is pulled through, the new thread is in place for subsequent dilatations. When the stricture appears to be well dilated and the child is taking all food by mouth, the thread is still kept in the esophagus for some weeks until it is apparent that stricture will not recur.

Surgical repair of atresia of the esophagus with tracheo-esophageal fistula is difficult technically, and postoperative care is constantly demanding. Correction of such a deformity in a four-pound infant is a test of delicate skill. The operation, far from simple and requiring from one and one-half to two hours, performed on infants often subnormal and weakened by pneumonia and/or atelectasis, is bound to be hazardous. When to these handicaps are added other severe abnormalities, especially of heart, kidneys and intestine—20 per cent of our cases had such multiple anomalies—it is understandable that hospital mortality will continue in the neighborhood of 30 to 50 per cent.

Congenital atresia of the esophagus *without* communication with the air passage constitutes about 5 to 7 per cent of cases Absence of air in the gastrointestinal tract suggests but is not diagnostic of uncomplicated atresia. We have had two infants whose roentgen film showed atresia of the upper end of the esophagus no air in the gastrointestinal tract, and yet had a tracheo-esophageal fistula The esophageal attachment to the trachea was small but adequate for a typical repair as outlined above These two cases confirm our opinion that the child with simple atresia of the esophagus should be explored transpleurally In our series of approximately 150 cases we have had about 10 with uncomplicated atresia In only one were we able to bring the ends of the esophagus together That child was a mongolian idiot and died the day after operation Pulling the stomach up through the diaphragm to furnish extra length to the lower end of the esophagus has met with failure It is worth while to explore these children with the hope of making an anastomosis but the parents should be warned before operation that the chances of its accomplishment are dim

The only possible procedure therefore is to bring the upper end of the esophagus out through an incision above and parallel with the left clavicle. The end of the esophagus will have to be drawn well out of the neck and carefully sutured to the skin The chest wound is closed and if the baby is in good condition, he is turned on his back and a gastrostomy is performed through a small incision in the left upper quadrant All fancy and complicated methods of doing a gastrostomy are out of order because the stomach is too small. The stomach is simply grasped with a couple of Allis forceps and opened by a stab wound A dePezzer catheter is inserted and encircled by a purse string of surgical gut in the stomach wall. With a few interrupted sutures the stomach is anchored to the peritoneum and fascia. Fascia must be included in the stitches or the stomach may pull away from the abdominal wall, allowing a catastrophic intra abdominal leak. When the wound is completely closed the catheter is drawn up snugly to hold more securely the stomach against the abdominal wall It is fixed with a few strips of adhesive tape on the skin.

What to do next with these children? Nothing much but routine care until the child is about two years old During this time the gastrostomy will require frequent toilet and regular changes of the tube A baby loves to suck on anything He is given an ordinary pacifier not only to afford some pleasure but also to keep him quiet. As soon as he is able to appreciate flavors he is given tiny amounts of food by nipple or teaspoon Of course the material runs right out of the esophagus but the flavors and the act of swallowing are educational One child who had not enjoyed these privileges had construction of an esophagus at two years of age and, because he had never learned the pleasures of eating, was a feeding problem for months

regurgitation was followed by aspiration into the lungs and consequent respiratory obstruction. Eventually—but too eventually—we learned to feed these infants slowly with a medicine dropper. It is now routine for the nurse to hold the infant on her lap for feedings. The medicine glass containing the formula is placed on a stand. The nurse fills the medicine dropper and gives it to the child. The time it takes the nurse to reach over and again fill the medicine dropper is sufficient to allow the previous amount given to pass through the esophagus. As a further safeguard an aspirating machine with catheter and suction tip, all connected, is ready for use in case the baby regurgitates or coughs. This simple process of slow feeding has reduced postoperative pulmonary complications and lowered the mortality rate remarkably. Such feeding is continued until it is demonstrated that the baby is ready for bottle and nipple feeding.

Upon discharge from the hospital the parents are instructed to note and report any difficulty in swallowing and are asked to return at three months for barium studies of the esophagus. After uncomplicated healing of the repaired esophagus stricture rarely occurs and dilatations are seldom necessary.

Gastrostomy is performed *only* if a sizable leak occurs at the anastomotic site in the esophagus. The gastrostomy then is necessary for feeding and later is advantageous for dilatations of the almost inevitable stricture. Before the stricture has become all but occlusive preparations are made for dilatations. A long piece of no. 2 braided silk is tied to a fine catheter which is guided through the esophagus into the stomach under fluoroscopic control. The catheter is fished out of the stomach through the gastrostomy, and the thread is pulled through. The ends of the thread are tied together. Dilatations are performed in the following way. The thread is cut, and the end near the gastrostomy is tied to a bougie. To the other end of the bougie a new thread is tied. By grasping the end of the thread near the mouth the bougie is pulled in retrograde fashion through the stricture and out of the mouth. As the bougie is pulled through, the new thread is in place for subsequent dilatations. When the stricture appears to be well dilated and the child is taking all food by mouth, the thread is still kept in the esophagus for some weeks until it is apparent that stricture will not recur.

Surgical repair of atresia of the esophagus with tracheo-esophageal fistula is difficult technically, and postoperative care is constantly demanding. Correction of such a deformity in a four-pound infant is a test of delicate skill. The operation, far from simple and requiring from one and one-half to two hours, performed on infants often subnormal and weakened by pneumonia and/or atelectasis, is bound to be hazardous. When to these handicaps are added other severe abnormalities, especially of heart, kidneys and intestine—20 per cent of our cases had such multiple anomalies—it is understandable that hospital mortality will continue in the neighborhood of 30 to 50 per cent.

at the anastomotic site in the neck At the first suggestion of narrowing dilatation should be begun

The most desirable method of constructing a food passage for those unfortunate children born with atresia of the esophagus will not be known until many long term records have been studied and analyzed during the next ten to fifteen years To this all will ascribe No child should be condemned forever to an open esophagostomy at the neck and a permanent gastrostomy

Congenital tracheo-esophageal fistula without atresia of the esophagus has been encountered only twice In one instance it was found at postmortem examination as one of many congenital anomalies In the other case it was diagnosed roentgenographically and successfully repaired. The gratification afforded by this operative triumph was somewhat dulled when the child at two months of age showed unmistakable evidence of gross mental deficiency

All over the world innumerable children born with atresia of the esophagus with or without tracheo-esophageal fistula now survive operative correction grow up and live normal, happy lives They owe a deep debt of gratitude to many workers but especially to the pioneers Logan Leven, William E. Ladd and Cameron Haight

Construction of a substitute esophagus, we believe, should be delayed until the child is about two years of age The structures then are large enough for safer handling, the blood vessels are more readily identified and less subject to compression, and the inlet to the thorax at the neck is large enough to accommodate more easily the segment of bowel to be used as the new esophagus

Stomach, jejunum or colon may be used to substitute for the missing esophagus Bringing the fundus of the stomach high into the chest and anastomosing it to the esophagus appears to be the least desirable method A portion of the clavicle and first rib have to be resected to make room for the stomach drawn up in the neck to meet the upper end of the esophagus, immediately after operation dilatation of the transplanted stomach occasions respiratory problems by encroaching upon air space

We have used the jejunum successfully as a substitute esophagus in three cases A Roux-en-Y operation was performed on the jejunum The single long arm of bowel was drawn behind the sternum and anastomosed to the upper end of the esophagus without technical difficulty The lower end of the jejunum was not anastomosed to the stomach because we feared gastrojejunal ulceration Although these children have gone three, five and six years without trouble and have maintained satisfactory growth and nutrition, we do not like the idea of bypassing the stomach

During the past two years a substitute esophagus has been constructed from the ascending colon in seven cases The appendix was removed, the ileum cut short and turned in, the ileocecal vessels severed, and the cecum freed and drawn up behind the sternum into the neck, where it was anastomosed to the esophagus The colon was cut near the hepatic flexure and anastomosed to the stomach It is claimed that the colon is more resistant to ulceration by gastric juice than the jejunum Intestinal continuity, of course, was re-established by an ileotransverse colostomy The operation was performed on one child who was only eighteen months old That was a mistake Insufficient room for the cecum at the thoracic inlet caused interference with the blood supply and resulted in a leak During a later attempt at repair of this fistula the anesthetist lost control, and the child died of hypoxia—commonly labelled cardiac arrest The other six have done well

We are inclined to believe from our meagre experience that the colon serves best for the construction of an esophagus Placement of the colon beneath the skin *in front of* the sternum is unjustifiable because of unsightly appearance Who wants visible peristalsis and bowel rumblings right beneath his shirt?

In each instance the gastrostomy tube was left in the stomach until fluoroscopic studies with barium demonstrated that the newly constructed esophagus was functioning well If a stricture develops, it is apt to be

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Respiratory Problems of the Newborn, of Infants and of Children*

The newborn child separated from its parasitic existence and ejected into an individualistic world suddenly is dependent upon its own respiratory system The lungs solid and full of fluid to the moment of birth, must promptly expand and begin the process of respiration It is little wonder that the few minutes immediately after birth are the most hazardous period in a person's life Any abnormality from mouth to diaphragm

* Portions of this chapter are taken from the Lord Moynihan Lecture, delivered before the Royal College of Surgeons and the British Association of Pediatric Surgeons, July 25 1958 and published in the Annals of the Royal College of Surgeons of England, 23 273 November 1958.

accentuates the infant's difficulty in accommodating itself to the new responsibility of breathing.

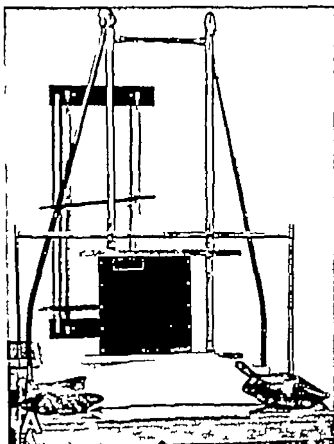
For diagnostic purposes the newborn infant with respiratory difficulties is promptly placed in one of two groups: the cyanotic without respiratory distress, or the dyspneic with or without cyanosis. The former group of persistently cyanotic infants usually has either congenital heart disease or brain injury. The surgeon's interest is focused upon the second group of infants, who are simply dyspneic or so severely dyspneic that they are also cyanotic.

Observation of the child's breathing is of first importance in deciding whether respiratory distress is due to pharyngeal, laryngeal or tracheobronchial obstruction, or to inadequate pulmonary function. Stertorous, coarse, rasping respiration indicates high obstruction due to such conditions as lingual cysts, micrognathia (Pierre-Robbins syndrome), macroglossia, choanal atresia, supraglottic webs, pharyngeal paralysis, and so forth. Laryngeal obstruction, far more common, is signalled by rapid, labored breathing, inspiratory stridor and an abnormal or absent cry. Obstructions below the larynx in the trachea cause inspiratory crowing or expiratory wheezing, but no interference with the normal cry. Inadequate pulmonary function manifests itself by rapid but not labored respiration.

Although observation of the child in respiratory distress and routine physical examination are important, nothing gives more direct information than good roentgenograms, which, to be of value, preferably are made in the x-ray department. Portable films are usually valueless and often misleading. It is always possible to move the infant to the x-ray department in an Isolette—in oxygen if necessary. By utilizing the apparatus shown in the illustration, clear anteroposterior and lateral films can be made with an exposure of 1/30 to 1/60 second. By holding the infant up by his arms, the scapulae are made to move laterally and the diaphragm is depressed. Films taken in the upright position are more satisfactory than in the supine because they are more quickly made, are free from the shadows of waving arms and legs and allow inclusion of an undistorted view of the abdomen. If the infant is in such severe respiratory distress that removal from the oxygenated Isolette seems hazardous, an attendant may direct with a funnel or cone a stream of oxygen over the baby's face during those few moments required for exposure of the films.

It seems appropriate rather dictatorially to state that, except for aspiration of the pharynx, no active surgical treatment is permissible until good x-ray films have been seen.

Responsibility for the care of respiratory emergencies in the newborn is shared by the endoscopist, the pediatrician and the surgeon. The majority of cases involving various parts of the respiratory passages are





handled by the endoscopist—at the Children's Memorial Hospital, Dr Paul Holinger and his associates To the surgeons fall the problems within the chest.

Congenital Diaphragmatic Hernia

A congenital diaphragmatic hernia in a newborn infant calls for prompt surgical intervention Even though the baby has relatively few symptoms, there is no excuse for the attitude, "Let's wait and see how the baby gets along and operate when he is a little older" Sudden distention of the bowels may quickly lead to acute respiratory distress and death

DIAGNOSIS Rapid respiration, cyanosis in the severely dyspneic, poor expansion of the chest on the affected side and a scaphoid abdomen suggest diaphragmatic hernia Percussion of the chest will yield inconclusive results because the note may be tympanitic or dull, depending upon the amount of air in the bowel Auscultation in a baby breathing sixty to seventy times a minute furnishes little information. Breath sounds are absent over the involved side and often diminished on the opposite side because of displacement of the mediastinum Bowel sounds in the chest, commonly heard in the adult, are rarely discernible in an infant

A barium swallow should not be given because it is not needed for a definitive diagnosis and is apt to be regurgitated and aspirated In the x-ray film a typical bowel pattern will be seen in one side of the chest and the heart pushed over to the opposite side Configuration of air shadows in the abdominal cavity will demonstrate what portion of the gastrointestinal tract remains in the abdomen

Differential diagnosis demands consideration of only one condition —postpneumonic lung cysts Infants may contract staphylococcal pneumonia early in life, and the cysts which sometimes form in the infected lung may be mistaken for loops of bowel A ten-day-old dyspneic infant, with no fever and with diminished breath sounds in the left side of the chest had what appeared from the x-ray films to be loops of bowel in the left chest In the lateral view we thought we saw loops of bowel going through a hole in the diaphragm At operation the diaphragm was painfully intact The child survived, although later drainage of an empyema was necessary In retrospective review of the history and findings one significant fact was apparent and should have impressed us more deeply—the white blood cell count was 25,000 per cubic millimeter This case, included in a published article on lung cysts, brought letters from two humiliated surgeons, each of whom wrote, "It was comforting to learn that someone else had made the same mistake"

PATHOLOGY. The majority of congenital diaphragmatic hernias, approximately 70 per cent, occur through a posterolateral defect in the

left leaf of the diaphragm and about 20 per cent through the right leaf The lesion in practically all cases consists simply of a hole of varying size in the diaphragm without a limiting peritoneal sac. In the few instances in which a sac persists extrusion of the bowels into the chest is limited. It seems more appropriate to label the case with a confining sac eventration of the diaphragm even though the wall of the sac consists only of fused layers of peritoneum and parietal pleura

Hiatus hernias in our hospital comprise not more than 5 per cent of cases Why they should be commonly seen in England and rarely in this country is unexplained The reason is not that we have not looked for them

The remaining 5 per cent of hernias occur through the foramen of Morgagni, an opening immediately posterior to the sternum

In only one instance have we seen bilateral congenital diaphragmatic hernia.

TREATMENT A number of years ago an infant with mild respiratory distress due to a congenital diaphragmatic hernia was admitted to the hospital shortly after midnight. It was decided that operation could safely be delayed until an open hour in the schedule at ten o clock in the morning. Suddenly about nine o clock, the infant became deeply cyanotic and appeared to be dying. He was snatched up and rushed to the operating room The abdomen was prepared with a quick slosh of iodine and hastily opened The bowels were withdrawn from the chest while the anesthetist with a face fitting mask forced oxygen into the lungs Slowly the pulse came back, the child s color improved and the operation was successfully completed in an orderly fashion Since then our policy has been to operate upon an infant with a diaphragmatic hernia when the diagnosis is made

On the operating table before anesthesia is begun a no 16 catheter is slipped through the mouth into the stomach to remove any accumulated fluid or formula. Intratracheal anesthesia is used. After a polyethylene catheter has been placed in the saphenous vein at the ankle a high left paramedian or subcostal transverse incision is made The bowels are extracted from the chest by grasping loop after loop with a layer of gauze between the fingers It is essential to insert a catheter through the hernial orifice to allow air to enter the chest as the bowels are being pulled down, or the vacuum will retract one loop as fast as another is withdrawn The bowels are wrapped in wet saline packs The hernial opening is retracted a bit to allow observation of the lung If the entire chest cavity on one side was completely filled with bowels the lung will be a nubbin no larger than the end of one s thumb and will expand very little as the anesthetist squeezes on the breathing bag.

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all have been properly placed As the last stitch is being tied as much air as possible is withdrawn from the chest cavity After the opening has been securely closed the peritoneal edges are drawn together with a running suture of fine surgical gut If the posterior rim of the diaphragm is missing, the anterior edge will have to be sewn to the rib edges posteriorly

Placement of the bowels in the abdomen is often difficult A helpful maneuver is to open a surgical sponge and lay it over the intestines The edges of the gauze are then tucked into the abdomen It is relatively simple to replace the bowels enclosed in this envelope of gauze mesh and compress them while closing the peritoneum with a running catgut suture This gauze tent is left in place and retracted to prevent loops of bowel from constantly popping out of the wound just as a stitch is being placed Before the last few sutures are put in, the gauze envelope is easily withdrawn The fascia and skin are closed with interrupted silk sutures Occasionally, in an infant who has all abdominal viscera but the liver in the chest, the undeveloped abdominal cavity will not accommodate the bowels and allow closure of the fascia The skin in such cases is freed and closed over the bowels with many interrupted silk sutures In the course of one week intra-abdominal tension will have stretched the muscles sufficiently to allow proper closure The second operation should not be delayed beyond the seventh day, or beginning fibrous adhesions between the bowels and the under surface of the skin will make separation difficult

While the child is still on the operating table as much air as possible is aspirated from the chest cavity It is well to insert the needle through the chest wall at an angle to avoid puncture of the expanding lung

Under no circumstances may the anesthetist put more than 15 cm of water pressure on the anesthesia bag in an attempt to expand the lung In two cases excessive pressure on the bag caused rupture of the opposite lung and consequent pneumothorax Leave the undeveloped lobe of lung alone, in the course of a few weeks it will expand spontaneously to fill the chest

A plastic suction tube is inserted through a nostril into the stomach, and the infant is returned to his oxygenated crib Negative suction is maintained on the gastric tube for two days to prevent distention and strain on the suture line Feedings are cautiously begun

Some surgeons prefer a transthoracic approach for repair of congenital diaphragmatic hernias The only time I tried this approach on the left side it was impossible to push the loops of intestine into the unaccommodating abdomen faster than they slid out, it was like trying to stuff foam into a bottle with a tweezer In despair the chest wound was closed, the abdomen opened and the hernia satisfactorily repaired

A transthoracic approach is appropriate for certain hernias through the right leaf of the diaphragm for cases in which only the stomach or a few loops of bowel are in the chest and for cases of eventration of the diaphragm

Postoperative care consists primarily in watching for and treating respiratory emergencies Results of surgical treatment of typical congenital diaphragmatic hernias are satisfactory and recurrences are few Mortality following operation is low in infants who at birth are near normal weight and have no other serious congenital anomalies

Pneumothorax

Pneumothorax in the newborn may be spontaneous or induced Atelectatic lungs full of fluid suddenly begin to expand at the moment of birth. It is logical to assume that too rapid expansion of one portion of the lung may lead to overdistention and that a sudden lusty cry may cause rupture of an alveolus Induced pneumothorax may be the result of too vigorous attempts at resuscitation Blowing in the infant's mouth is still a common practice and, if too energetically applied, may lead to rupture of the lung It is natural for the obstetrician to become alarmed when an infant does not breathe and is cyanotic, and in desperation to apply his own full lung pressure to the baby's mouth to induce adequate respiration

Pneumothorax may be confined to the mediastinum in those cases in which an alveolus ruptures beneath the visceral pleura. Air seeps through ruptured alveoli and finds its way proximally along blood vessels to the mediastinum. Large blebs and accumulations of air behind the sternum may be sufficiently large to interfere with return of blood to the heart by the mechanism referred to as the "air block syndrome"

Roentgenograms will show air shadows about the heart and large vessels What needs to be done is purely a matter of circumstance and judgment often no treatment is required If respiration is impeded to the point that relief is necessary air may be aspirated by needle or a thoracotomy may be necessary for puncture of offending cysts Just what to do and how much to do at what moment are matters of individualization for each patient.

Typical pneumothorax with collapse of a lung due to rupture of an alveolus through the visceral pleura into a chest cavity is more common than mediastinal emphysema and more easily handled. A good roentgenogram makes the diagnosis Needle aspiration of trapped air is often lifesaving The impulse to leave a needle in the chest and attach it to suction must be denied because the expanding lung may hit the needle point and be further injured. After removing the free air from the chest with a syringe and needle attached to a two-way stopcock, it is safe to

wait a bit to determine whether air will reaccumulate if it doesn't, nothing further is required, if it does, a small catheter is introduced into the chest between the ribs and attached to gentle negative suction to re-expand the collapsed lung A continuous stream of air flowing into the suction bottle for a number of days should suggest a congenitally open bronchus We overlooked this possibility in an infant and at postmortem examination found just that—an open bronchus in the middle lobe which could easily have been removed An unexplained, persistent pneumothorax in the left side of the chest should arouse suspicion of a ruptured esophagus, a diagnosis easily confirmed or disproved by a swallow of Lipiodol

Congenital Lung Cysts

Congenital lung cysts in infants are of special interest because they are uncommon and yield dramatically to surgical treatment Not at birth, but usually between the fifth and tenth days of life the infant has increasing dyspnea and eventually cyanosis Roentgenograms are diagnostic In acute cases the expanding cyst fills one side of the chest and pushes the mediastinal structures to the opposite side

Having made a diagnosis of a progressively expanding lung cyst, it is essential that the surgeon with a 50-cc syringe and needle in hand remain with the child while the operating room is being prepared for emergency surgery. If at any moment dyspnea becomes alarming, the cyst is aspirated It is unwise after relieving dyspnea by aspiration to leave the child for even a moment, because the cyst may rapidly refill with air or, if the punctured cyst collapses, an equally dangerous pneumothorax may result The surgeon accompanies the child to the operating room

Operative removal of the lobe or section of the lung from which the cyst arises is easily accomplished Our experience with ten cases has been happy because of good fortune in having the cyst in all instances confined to one lobe of the lung Generalized cysts involving an entire lung leave the surgeon no choice but to perform a pneumonectomy, although recognizing that infants tolerate this procedure poorly

Congenital lung cysts may be encountered also in children and teenagers Recurrent accumulations of pus in the same area in the chest and usually treated as empyemas should suggest the possibility of a lung cyst as the basic cause A lung cyst may be an incidental roentgenographic finding On the basis of clinical findings and roentgenographic findings it is often difficult to be sure whether a lung cyst is congenital or postpneumonic (see next section) A definitive diagnosis of congenital lung cyst is made on microscopic evidence of smooth

muscle and/or cartilage in the wall of a cyst lined by respiratory epithelium

Postpneumonic Lung Cysts

Pneumococcal and streptococcal pneumonias in infants and children respond so promptly to antibiotics that the surgeon sees practically no complications from these infections On the other hand, staphylococcal pneumonia, especially during the first year of infancy has become more common during recent years and, as the Staphylococcus has acquired resistance to antibiotics complications have increased, the most common is postpneumonic lung cyst or pneumatocele

PATHOLOGY A small, localized area of infection occurs in the wall of a bronchiole. The consequent inflammatory swelling completely or partially obstructs the air passage Complete obstruction causes local atelectasis but incomplete obstruction, because inspiration is more forceful than expiration leads to ballooning of the distal segment of lung or the formation of a postpneumonic lung cyst.

COURSE. The course of the disease is somewhat as follows The infant contracts bronchopneumonia which is unaccompanied by the classic signs and symptoms so vividly recorded in Osler's Practice of Medicine but with cough, fever and signs of mild or moderate toxemia In roentgenograms of the chest of infants ill enough to be hospitalized varying degrees of localized or diffuse congestion are seen. After a few days—no specific period of time—a second roentgenogram may show one or more round translucent shadows (The percentage of cases of staphylococcal pneumonia complicated by cyst formation is difficult to estimate—probably not more than 5 or 10 per cent of cases) The course of these postpneumonic cysts or pneumatoceles is tremendously variable (*a*) A single cyst or a number of cysts each a centimeter more or less in diameter may disappear in a week or two (*b*) The cyst or cysts, occupying as much as half the space in one side of the chest, may persist for months and then spontaneously disappear (*c*) Accumulation of pus in a cyst may occur it is easily recognized by evidence of a fluid level in the x ray film (*d*) A rapidly expanding cyst may rupture and produce acute pneumothorax and empyema.

TREATMENT The watchword in the treatment of postpneumonic lung cysts has been borrowed from the obstetricians—"watchful waiting" Even though the cyst enlarges and persists and the medical staff urges surgical intervention, the treatment is still intelligent observation Exceptions arise only when the cyst ruptures, and then prompt surgical intervention is necessary Those infants who are rather ill and have large pneumatoceles are watched for respiratory distress Provisions are made for emergency aspiration of the chest by keeping a wrapped sterile

syringe and needle at the crib side ready for use in case the cyst should unexpectedly rupture and produce pneumothorax. Hasty aspiration of the chest will relieve the acute respiratory distress, but insertion of a catheter into the chest cavity and mild negative suction invariably will be necessary to re-expand the lung and to remove pus which, if not already present, will almost surely form.

During the past twelve years forty-eight patients with lung cysts following pneumonia have been seen at the Children's Memorial Hospital. Two deaths occurred, one was due to pneumothorax observed by a member of the house staff who, because of no previous experience, failed to realize the need for quick action, the other was due to overwhelming pyopneumothorax and septicemia.

The surgical staff at our hospital succumbed to the demand for operative removal of postpneumonic lung cysts in only one case. An acutely ill, seven-month-old infant was hospitalized because of fever, cough and dyspnea of six days' duration. X-ray films confirmed a diagnosis of bronchopneumonia complicated by a number of pneumatoceles which occupied two thirds of the right chest cavity. The bronchopneumonia cleared, but the cysts, checked repeatedly by roentgenograms, persisted for six months. Because the cysts remained so long unchanged, the radiologist, Dr. Harvey White, claimed that the cysts were of congenital origin. The surgical staff stubbornly labelled them postpneumonic. Unfortunately, no roentgenograms previous to the acute illness were available to answer the question. Because of mild dyspnea—questionable—we yielded and resected the right lower lobe, which was indeed cystic. At operation no cysts could be found in the middle or upper lobes, but a few days after operation two cysts, each about 2 to 3 cm. in diameter, were demonstrated roentgenographically in the middle or right upper lobe. In the meantime the pathologist, Dr. Joseph Boggs, studied the specimen and made a diagnosis of congenital lung cysts. A later x-ray film showed that the two remaining cysts had disappeared spontaneously. The final diagnosis recorded on the medical record is "congenital lung cysts." The surgeons, still unconvinced, meekly paid off their small wagers in the snack bar. The child is well.

Congenital Lobar Emphysema

Congenital lobar emphysema is a relatively rare but specific cause of slowly increasing respiratory distress in infants less than six months old at the time of onset of symptoms. The emphysema, usually affecting one of the upper lobes, is due to congenital malformations of the cartilaginous rings in the bronchial wall, the rings may be flattened or stenotic, or small segments may be missing. The constricted or collapsible bronchus allows forceful inspiration, but not passive expiration. In-

creasing amounts of air become trapped in the lobe until overdistention encroaches upon the rest of the lung and produces respiratory distress

The only symptom slowly increasing dyspnea, may appear at the end of the neonatal period, but rarely becomes severe enough to be classed as urgent for a number of months The infant eats poorly gains weight slowly and even at rest breathes rapidly After bouts of crying dyspnea is notable The percussion note on the involved side of the chest is tympanitic the heart and mediastinal structures are pushed to the opposite side Roentgenograms of the chest are diagnostic and resemble those of congenital lung cysts in every respect except for this specific difference the dim outline of the structure of the lung can be seen throughout the tremendously distended emphysematous lobe

The treatment is surgical Upon opening the chest, the emphysematous lobe occupying most of the right or left chest cavity literally herniates itself out of the wound. The surface of the involved lobe is dotted with innumerable, tiny emphysematous blebs The lower lobe is compressed into a mass no larger than the end of one s thumb After the affected lobe has been removed the anesthesiologist, by making slight pressure on the anesthesia bag, can partially inflate the lower lobe The chest is drained for forty-eight hours by a catheter attached to under water seal drainage Within a couple of weeks the lower lobe will have completely filled the chest cavity

Our experience with this unusual condition is limited to nine cases All patients have remained well and symptomless since operation An upper lobe was involved in eight cases the right middle lobe in one

Congenital Bronchial Cysts

Congenital bronchial cysts may arise from either a bronchus or from the trachea They are thin-walled, contain mucoid material and are lined with ciliated columnar epithelium. Such cysts are uncommon we have operated upon only three Contrary to reports in medical literature, all our cases occurred in infants and caused moderately severe respiratory distress. In the x ray films a well outlined radiopaque oval mass could be seen compressing the trachea in one instance and a bronchus in two cases. Although successful surgical removal of the cysts was accomplished in all an interesting problem of technique presented itself in the first case in which a cyst was removed from the trachea. The cyst measuring approximately 3 by 6 cm., was easily freed except at its point of origin from the trachea. During the final dissection the thin fragile cyst was broken at its attachment to the trachea and a defect about 2 by 4 mm in the tracheal wall was uncovered What to do? Closure of the hole by suture would undoubtedly lead to tracheal stenosis An observer who had just come from Johns Hopkins University

Medical School said that he had heard of some experimental work there on closure of defects in the trachea or bronchus with a piece of Gelfoam The idea seemed sound A tiny patch of compressed Gelfoam was laid over the hole and fixed to the wall with a few interrupted sutures of 6-0 silk A flap of pleura was then drawn over the patch and sutured To my surprise, the trachea healed without postoperative leak and without subsequent stricture The same technique was successfully used to close a similar defect in a bronchus

Vascular Ring, or Anomalies of the Aortic Arch

In considering the complexity of embryologic development of the great vessels, it is little wonder that anomalies of the aortic arch appear in diverse forms The most common anomaly described in medical literature is that of persistent double aortic arch, in which one segment passes in front of the trachea, the other behind the esophagus One segment is always dominant and carries most of the blood In our surgical experience with forty-one anomalies of the aortic arch the most common variation from normal has been this The aortic arch deviates to the right Immediately beyond the arch an anomalous vessel arises from the descending aorta and courses to the left behind the esophagus, and turns slightly anterior where, as the ductus arteriosus, it is attached to the main pulmonary artery From the posterior placed anomalous vessel the left subclavian artery arises The left carotid artery arises from the ascending aorta and lies anterior to the trachea Often the left carotid and left subclavian communicate with each other immediately to the left side of the trachea This arrangement of vessels does not form as tight a ring about the trachea and esophagus as the typical double aortic arch A third relatively common and usually symptomless anomaly is that in which the right subclavian artery arises from the descending aorta on the left and passes to the right behind the esophagus Numerous less common anomalies of the aortic arch are seen, all vary in the degree in which they compress the trachea and esophagus from no obstruction whatsoever to severe, suffocating occlusion

DIAGNOSIS An infant who has labored respiration with suprasternal and infrasternal retractions, who constantly holds his head far backward, who feeds poorly and during feedings is apt to have attacks of cyanosis and syncope, most likely has an obstructing vascular ring A similar diagnosis is in order in the two- or three-year-old child who has had frequent upper respiratory tract infections associated with inspiratory stridor, and who constantly has a peculiar cough which resembles the bark of a sea lion

The diagnosis is confirmed by giving the child, placed in the lateral position before the fluoroscopic screen, a swallow of contrast medium—

Lipiodol to an infant, barium to a child—and observing an indentation in the posterior wall of the esophagus A good roentgenogram will show narrowing of the trachea at the same level If suitable visualization of the esophagus is impossible because the child will not swallow the contrast medium, an injection of a few centimeters of Lipiodol—warmed to promote easy flowing—through a catheter introduced through the nose or mouth into the upper end of the esophagus will be effective in demonstrating the deformity At endoscopic examination a pulsating vessel may be seen at the site of indentation of the esophagus

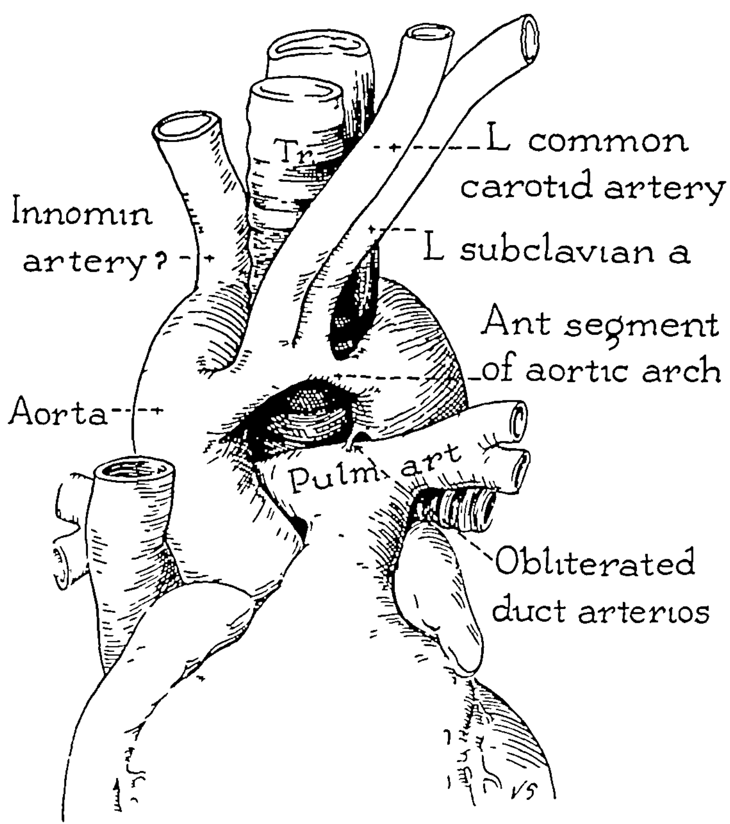
TREATMENT If symptoms warrant, operative treatment is indicated

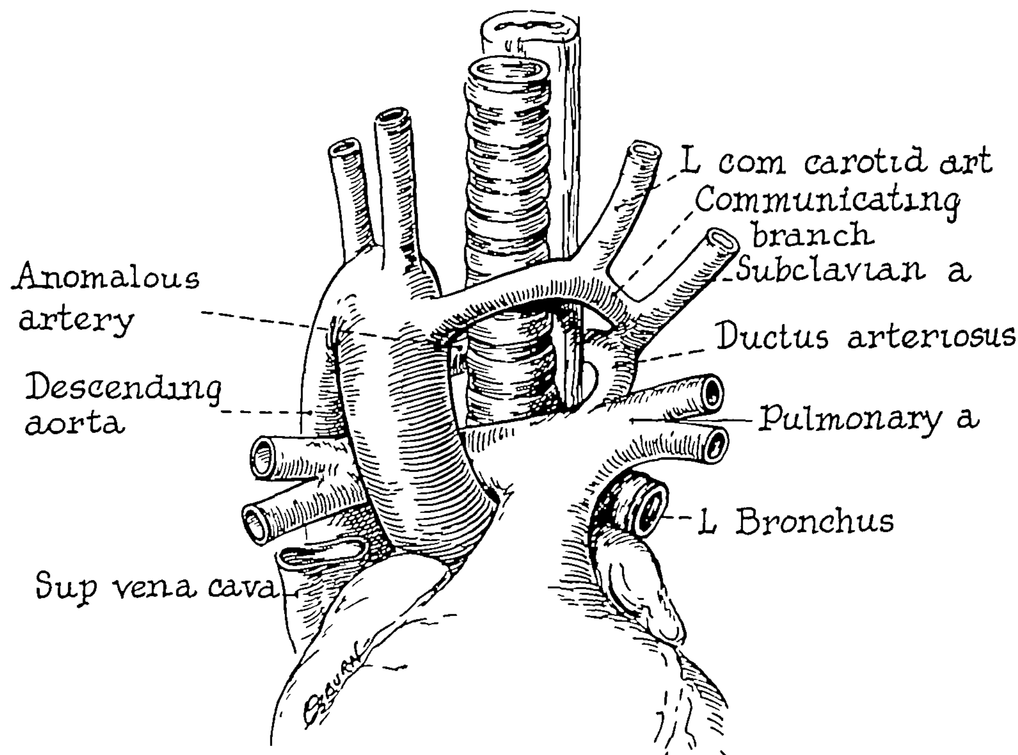
All anomalies of the aortic arch are approached under intratracheal anesthesia through a left submammary incision. The chest is opened through the third intercostal space and to obtain adequate exposure, the second and third ribs are cut at the costosternal junction Other surgical approaches have been tried and found wanting. While carefully avoiding injury to the phrenic and recurrent laryngeal nerves the great vessels at the base of the heart are thoroughly exposed and identified Before cutting any vessel it is well momentarily to occlude and release it while the anesthesiologist confirms identification by feeling the carotid and brachial pulses

In the typical case of double aortic arch the smaller vessel, anterior or posterior is clamped, cut and sutured. A small posterior constricting segment of a double aortic arch may be missed because no abnormality can be seen until an incision is made in the posterior parietal pleura and the area behind the esophagus is explored The first patient I operated upon for vascular ring in 1946 was a four week-old, extremely dyspneic infant. At operation no anomaly could be found. Never having heard or read of a small, persistent posterior aortic arch, it did not occur to me to open the posterior mediastinum. A few days after operation the child suddenly died. The postmortem finding of a small posterior arch, which could easily have been severed was difficult to take with equanimity

The case of right aortic arch with anomalous vessel behind the esophagus is corrected by dividing the ductus arteriosus the left subclavian artery and the connection if present, between the carotid and subclavian arteries The remainder of the anomalous vessel is then drawn to the left and securely ligated as near as possible to its origin from the descending aorta. The distal segment of the vessel is removed To relieve compression on the trachea, the left carotid artery is elevated and attached to the posterior surface of the sternum by a silk stitch which catches only the fibrous tissue about the vessel.

An anomalous right subclavian artery coming from an aorta descending on the left is commonly seen during cardiac surgery Only if


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artery
Ductus arteriosus
Descending
aorta
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Sup vena cava

symptoms of dyspnea or dysphagia have been noticed before operation is the vessel freed from its position behind the esophagus and cut

The greatest hazard in this operation is that of anesthesia. The site of tracheal compression just above the carina, is difficult to pass without pushing the tracheal tube into one or the other bronchus Placing the intratracheal tube proximal to the obstruction results in inadequate aeration and unsatisfactory anesthesia. Recently our anesthesiologists have devised and successfully used a small plastic intratracheal tube in the distal end of which a number of holes have been cut. The perforated end of the tube placed in the constricted portion of the trachea, allows dispersion of gases and adequate respiration

After operation, breathing is apt to be labored because of traumatic edema in the previously constricted area of the trachea. All children are placed in highly humidified and well oxygenated cribs and watched closely for signs of respiratory distress Tracheotomy is done, not as a last resort, but when it is apparent that respiration is inadequate Unless the tracheotomy tube extends beyond the point of obstruction little relief is obtained.

To relieve dyspnea following operation for a very tight vascular ring on a two-month-old infant a polyethylene tube was introduced through a tracheotomy tube through the site of inflammatory obstruction in the trachea and into the right bronchus where it was actually left for eleven days It did not seem possible that a tube in the bronchus could be tolerated for such a prolonged period but there was no alternative each earlier removal was followed by recurrence of severe dyspnea demanding its immediate replacement. The infant survived

Results from release of a tight vascular ring in infants are dramatic. Children who have had mild obstruction and have had a characteristic cough improve slowly after operation To avoid disappointment, it is well to warn parents that the odd sounding cough may persist for some months and may even later recur during respiratory infections

Bronchiectasis

ETIOLOGY Bronchiectasis in children is not only becoming less common each year but is also appearing in less severe forms Adequate treatment of bronchopneumonia has partially eliminated one of the chief etiologic factors Better food, better housing and better general care have aided in building up resistance to unresolved foci of infection in the lobules of the lungs—the starting point for bronchiectasis

Two etiologic factors are less well controlled a foreign body in a bronchus and mucoviscidosis Radiopaque foreign bodies are easily recognized and, if promptly removed cause no irreversible changes The trouble makers are such objects as peanuts, hay pine needles hard

peas—the kind youngsters use in pea shooters—and many small plastic and glass objects After inhalation of a foreign body the child chokes and coughs A persistent cough eventually prompts the parents to seek medical advice A roentgenogram, if taken, will show no foreign body, but will show a patch of congestion in one of the lower lobes of the lung, usually the right A history of unexplained sudden choking followed by persistent cough is sufficient reason for an x-ray film of the chest An area of congestion, whether a foreign body is visible or not, is an absolute indication for bronchoscopic examination The skill with which expert endoscopists can go into those tiny bronchi and pull out safety pins, thumb tacks, peanuts, bobby pins, heads of timothy hay, and so on, never ceases to be amazing and, to those who have never used a bronchoscope, miraculous

The time it takes a foreign body to produce permanent irreversible changes in the bronchioles depends upon the type of foreign body inhaled Vegetable foreign bodies (peanuts and peas) are the worst offenders

Mucoviscidosis produces bronchiectasis in much the same manner as foreign bodies Thick secretions block a bronchus and produce local atelectasis in which infection smolders A small abscess forms, and eventually a number of bronchioles become diseased and permanently damaged

Chronic nasal sinusitis has long been blamed as a cause of bronchiectasis Perhaps this accusation is just, but it seems to be a minor factor in children—all children's noses drip about one third of the time It is possible that chronic sinusitis in children persisting through the teen-age period and into adulthood eventually leads to cases of bronchiectasis seen in people between twenty and forty years of age There is also some evidence that bronchiectasis causes sinusitis

In our experience the pneumonia following whooping cough and measles seems more prone to lead to bronchiectasis than standard types of pneumonia This observation may be an impression rather than a bona fide statistical observation

TREATMENT Bronchographically demonstrated saccular and tubular—especially saccular—dilatation of a number of bronchioles in a persistently collapsed portion of lung is a positive indication for surgery The child who regularly coughs up foul-smelling sputum and has a persistent roentgenographic shadow in the lung indicative of smoldering infection is a candidate for surgery even though few changes are demonstrable in the bronchogram On the other hand, the child who has few general symptoms, some chronic cough, but little sputum except in the morning, and yet has considerable puddling of Lipiodol in the bronchioles of one lobe of lung is entitled to a vigorous regimen of medical management

Whenever indications for surgery are not absolute, medical man-

agement is in order General health is attended to and broad spectrum antibiotics are given for months The parents are taught what is meant by postural drainage and advised to apply it one or more times a day depending upon the amount of sputum brought up each time. Most sputum will be coughed up in the morning A demonstration of postural drainage *is given The child rests his legs and abdomen on a bed or sofa and leans forward at a 60-degree* angle A sputum cup is appropriately placed. The child is urged to cough and expectorate anything that comes into the throat. Gentle pounding on the back of the chest stimulates cough Children like the patting or pounding—it makes them feel that their parents are cooperating If the sputum lessens after a reasonable trial and becomes free of purulent material conservative treatment is certainly in order It is questionable whether bronchiectasis ever completely disappears but the fact of its existence alone is not an indication for surgery

A child is a candidate for operative treatment if after each cold a nasty, wracking cough persists some mucopurulent material is coughed up each day and repeated bronchograms show unchanging saccular bronchiectasis Operation should not be delayed because purulent secretions spilling over into other lobes of the lung will eventually compound the disease process

Children with fibrocystic disease who have extensive bronchiectatic changes confined to one lobe of the lung, although not curable, are greatly benefited by lobectomy

Infants and young children occasionally have atelectasis of the middle lobe, similar to the "middle lobe syndrome" described in adults If the lobe remains atelectatic in spite of repeated bronchoscopic aspiration and if Lipiodol injected into the lobe remains there for days lobectomy is indicated

Operation A day or two before operation for bronchiectasis bronchoscopic clearance of the lung is in order to minimize the danger of aspiration during anesthesia. Intratracheal anesthesia is given and as soon as the child is anesthetized the bronchi are again cleared with a long polyethylene suction tube

Examination and palpation of the exposed diseased lobe will frequently indicate a more extensive lesion than was anticipated from the bronchogram If the disease is confined to one lobe a simple lobectomy is routinely performed. If however the disease involves more than one lobe of lung, segmental resections are preferred For example a seven year-old girl had demonstrable bronchiectasis of the lower lobe and the lingula of the upper lobe on the left side and of the middle and lower lobes on the right There was a history of whooping cough and pneumonia at age two years and measles and pneumonia at age five At the first operation the entire middle lobe and two segments of the right lower lobe were removed. At the second operation the lingula and

involved segments of the left lower lobe were removed Five years after operation she has no subjective or objective signs of respiratory insufficiency

If the left lower lobe alone is involved, a lobectomy is done. In our experience, however, the lingula of the upper lobe is practically always involved and requires resection.

The technique of lobectomy is entirely by sharp dissection with a scissors First the artery is isolated, carefully identified, doubly tied and cut, then the veins are similarly handled The isolated bronchus is temporarily occluded while the anesthetist inflates the rest of the lung to demonstrate patency of the remaining air passages The bronchus is clamped with an angulated, toothed ductus forceps which will occlude but not crush the bronchial wall No bronchus should ever be crushed with the ordinary type of hemostat Crushed tissue will not heal as well as uncrushed tissue The bronchus is cut about 2 mm distal to the toothed clamp, the stump is sponged with Zephiran, closed with a proper number of 5-0 silk sutures on an atraumatic needle and covered with a bit of pleura "Pride goeth before destruction, and an haughty spirit before a fall" (quote from a reprint by Solomon) With these protective words as a charm against possible trouble next week, it is stated that in sixty-eight lobectomies, twenty-five for bronchiectasis, we have had no case of postoperative opening of a bronchus

Results following operation have been good, and mortality has been nil These facts do not alter our conservative attitude toward surgery for mild bronchiectasis Before operation is advised for the child with borderline symptoms and laboratory findings of bronchiectasis, the physician, parents and surgeon must be convinced that medical management has failed and that risk of surgery is justified

A word of warning from an almost burned hand A six-year-old boy who had been treated for pneumonia in February had an x-ray picture of his chest a month later because of persistent cough The left lower lobe of the lung was completely collapsed Six weeks later it was still collapsed In June a bronchogram was made, and tubular bronchiectasis was demonstrated in the persistently collapsed lung Mild symptoms persisted, and operation was advised, but, because of coming vacations, was delayed until August Some guiding power suggested that an x-ray film of the chest be taken the day before operation The lobe had completely expanded and was normally clear The child has remained well

Pectus Excavatum

Funnel chest, or pectus excavatum, is another one of the conditions in childhood about which little is known except that the deformity

occurs Etiology is obscure, indications for operation are equivocal, and methods of correction are assorted.

Indications for Operation Our attitude towards operative correction lies somewhere between that of operating upon the majority of children with a depressed sternum and operating upon none. Between these conflicting camps we have pitched our defenseless tent—in that section of the neutral zone which lies adjacent to the nonoperator's territory

Operation is not advised for pectus excavatum during infancy A number of years ago I made a small transverse incision over the xiphoid of a deeply depressed sternum in a six month-old infant, expecting to cut the substernal ligament. It was rather mortifying to be able to find no substernal ligament The only attachments to the sternum were the fibers of the rectus muscles and they appeared to be exerting anterior traction by reason of a protuberant abdomen That operation has not been repeated. Incidentally in a fair number of operations for pectus excavatum on older children no substernal ligament has even been seen.

Funnel chest is preferably corrected in children between the ages of two and five years Decisions for or against operation in this age group are made after physical and roentgen examination, and after discussion of psychologic factors

After the child with pectus excavatum has been disrobed a glance is usually sufficient to decide whether operation is or is not indicated. The child who has a mild to moderate depression of the sternum and slight protrusion of the abdomen does not need operation nor will the frequency of colds complained of by the parents be lessened by operation It then becomes the function of the examiner to convince the parents that operation is not indicated. By lifting the child by his arms and by bending him backward, it is demonstrated how the abdomen flattens and lessens the apparent degree of sternal depression Of course the surgeon listens to the child's heart and lungs, palpates the abdomen and on his more alert days even looks at the throat and examines the inguinal regions Roentgenograms of the chest in anteroposterior and lateral positions are made and shown to the parents to convince them that the heart is in its normal position and so forth. When the patient is a little girl, it is circumspectly explained that as she develops at puberty the slight depression of the sternum will in a measure accentuate her points of femininity

The child who has a deep depression of the sternum and a grossly protuberant abdomen, who is underweight undersize eats poorly and listlessly sits or stands in a slouched position is likely a candidate for operative correction He needs more breathing space. If roentgenograms of the chest show displacement of the heart to the left and flattening of its silhouette operation unquestionably is indicated

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the anterior surface of the chest with adhesive tape Parents are happier if the child wears this breastplate for a few weeks

Results in general have been satisfactory A few children have had considerable recurrence over a period of years The majority of children have a slightly flatter chest than normal, but not actually a recurrence

It has been surprising to observe the general changes in some of these children they are more active gain weight and instead of being quiet and reserved are vigorous and aggressive

There is little doubt that a few children require correction of funnel chest and that properly chosen patients will surprise the surgeon by their improved health.

Whether to advise operation for children who have extensive depressions of the sternum, but no abnormal symptoms, is a moot question When discussing pectus excavatum on ward rounds one day a resident told of a classmate who, in spite of a deep funnel chest, was a star swimmer at the University of Michigan and later made the Olympic swimming team This man was traced to Korea, from where he kindly sent photographs of his chest and x-ray films There was no question about the fact that he had rather deep depression of the sternum Who knows what swimming records he might have broken had he had the pectus excavatum corrected during childhood? When parents of a boy with a deep funnel chest, but otherwise quite well, were told that operation in his case would be largely cosmetic, they promptly put on their wraps and went home In another similar situation the father said that his brother had always been embarrassed because of a funnel chest and that he chose to have his son's deformity corrected It is apparent that the signs for operation are not clearly staked out

OPERATION. Intratracheal anesthesia is necessary for operative correction of pectus excavatum because the pleural cavity is often unavoidably opened A longitudinal incision is made over the sternum Some surgeons advise a transverse skin incision for the sake of better healing and less keloid formation We have tried the transverse incision, but find it awkward The pectoralis muscle attachments are separated from the anterior surface of the sternum

From this point on we have used and discarded a number of techniques and at present use the method popularized by Ravitch Linear incisions are made in the perichondrium of the five deformed costal cartilages on each side of the sternum By firmly grasping and pulling on the edges of the perichondrium it is possible with a thin periosteal elevator to peel the perichondrium from the cartilages The deformed cartilages are completely removed, and the flexible tubes of perichondrium are left intact Some patience and a little practice are necessary to accomplish removal of cartilages without tearing the perichondrium all to pieces

A transverse V-shaped incision is made in the anterior surface of the sternum at the level of the second intercostal interspace The sternum is hinged upward to its proper position, where it is held by coapting the edges of the V cut with a number of interrupted stout silk sutures For years we further supported the sternum with an encircling wire attached to a bar taped across the chest This wire is apparently unnecessary if the patient is below five years of age The pectoralis muscles are brought together at the midline and attached to the anterior surface of the sternum The skin is closed with silk The wound is sealed with adhesive tape over a gauze dressing To prevent the child from bashing in his poorly supported sternum, we fashion a plaster of paris slab and fix it to

Interatrial Septal Defects

So many articles recently have been written by expert cardiologists on the diagnosis of interatrial septal defects that any detailed review by me would be repetition—more honestly plagiarism. Excellent chapters on this subject are available in two recent books one by Nadas on "Pediatric Cardiology" the other by Keith, Rowe and Vlad on "Heart Lesions in Infancy and Childhood"

Diagnosis

The surgeon should be familiar with some of the fundamentals of diagnosis A child, usually above three years of age with an interatrial defect of the secundum type is referred to a cardiology center not be

defects is attended by the same low mortality rate as closure of a patent ductus arteriosus our indications for operation may be broadened

On the other hand the child who has moderate cardiomegaly and a large left to-right blood flow should be operated upon at the earliest convenient time even though no symptoms referable to the defect have manifested themselves

The child who is in heart failure because of a huge secundum defect or more commonly because of an ostium primum is a candidate for surgical correction after the heart has been restored to its best possible condition by digitalis and rest

Advanced pulmonary hypertension during childhood is not as com monly associated with interatrial defects as with interventricular defects but when present and equal to systemic pressure is of serious import operation upon such patients is attended with high mortality The fact that medical management has little to offer patients with severe pul monary hypertension probably justifies the high risk of mortality pro- vided a constant reverse shunt from right to left has not occurred

Surgical repair of interatrial septal defects began as a closed pro- cedure but because of frequently incomplete and often improper closure soon gave way to suture under direct vision through the opened atrium Today two methods are in vogue closure under hypothermia, or with the aid of extracorporeal circulation Each method has some attractive features Were it not for the fact that differential diagnosis between secundum and primum defects is not infallible and that all anomalous pulmonary venous communications with the atrium cannot rapidly be repaired, closure under hypothermia probably would be the operation of choice

In spite of these disadvantages of hypothermia, we continue to use it for closure of the secundum defects Only once because of an error in diagnosis was it necessary to close the chest and later reoperate to close the ostium primum with the aid of extracorporeal circulation

Our attitude at this moment is to advise open heart surgery under hypothermia for those patients only who without question have a small or moderate-sized ostium secundum defect. Open heart surgery with extracorporeal circulation is used for those children who have a large secundum defect with a huge left to-right shunt for those who have any evidence of anomalous pulmonary venous drainage and for those in whom there is the slightest suspicion that the defect may be an ostium primum.

Operation

For closure of the simple secundum defect the patient is anes thetized placed in ice and the temperature reduced to 85° F During

cause of ill health or decreased activity, but because of a heart murmur A moderately harsh blowing systolic murmur is best heard to the left of the sternum at the second or third interspace With a little practice a rumbling diastolic murmur can be heard over the apex in children with a large defect The pulmonary sound is accentuated, and the second sound is often widely split. A mild thrill is palpable in a small percentage of cases Enlargement of the right side of the heart and clouding of the lung fields as seen in roentgenograms are directly proportional to the size of the defect The electrocardiogram shows right bundle branch block and may show right ventricular hypertrophy These few findings indicate that the child has an ostium secundum defect

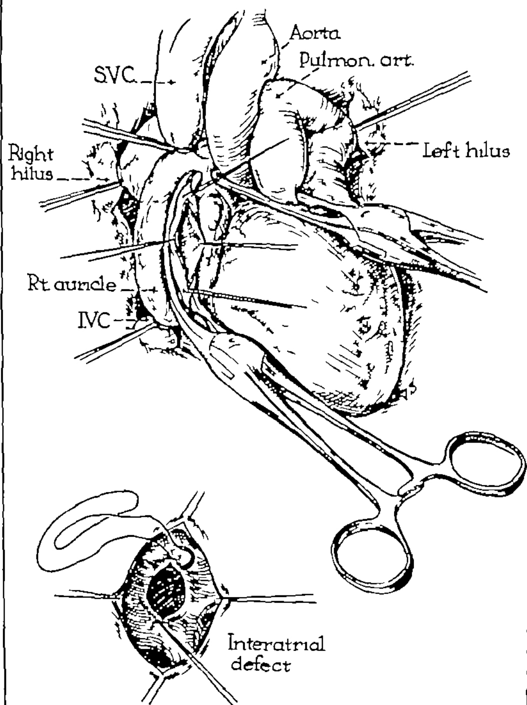
On the other hand, the child with an ostium primum defect, but especially with an atrioventricularis communis, is frequently referred to the cardiologist because of poor general health, debility and often a history of heart failure during the first two or three years of life Let's face it most cardiac surgeons are incapable of interpreting the fine points of clinical and laboratory studies for unquestionable differentiation of primum, secundum and atrioventricularis communis defects The pediatric cardiologist and the cardiophysiologist usually furnish the answer on the basis of history of onset of symptoms and their severity, size and shape of the heart, variations in cardiac impulse, fine characteristics of murmur, detailed electrocardiographic studies, cardiac catheterization data and selective angiocardiography As a matter of fact, in our surgical experience with the last thirty interatrial defects they made only one error in diagnosis—an ostium primum defect was mistaken for an ostium secundum

Partial anomalous pulmonary venous drainage is not uncommonly associated with an ostium secundum defect The extent of this anomaly too should be known before deciding what type of operation is to be performed

Indications for Operation

After completion of clinical and laboratory studies a conference by the entire department for evaluation of the case is educational as well as valuable for choice of proper operative procedure

At present we do not believe that operation is indicated for the symptomless child with a small secundum defect, provided the heart is normal in size or only questionably enlarged, and the pulmonary blood flow is less than twice that of the systemic flow It seems reasonable to delay operation until indications are definite enough to warrant the small hazard of surgery We do not at the moment agree with those who claim that the mere presence of an interatrial defect calls for operation the same as a patent ductus arteriosus When operative closure of interatrial


Aorta
Pulmon. art.
SVC
Left hilus
Right hilus
Rt. auricle
IVC
Interatrial defect

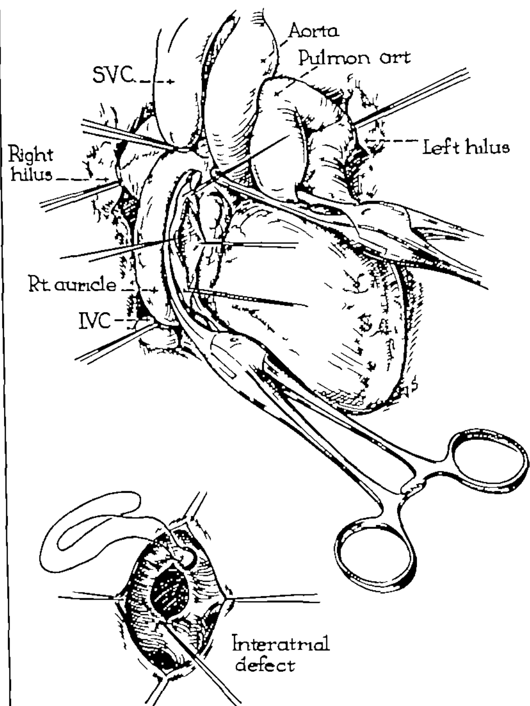
the cooling process continuous temperatures are recorded by two thermocouples—one in the esophagus, the other in the rectum. Two simultaneous readings are taken as a precaution against excessive hypothermia due to a faulty thermometer. During the early stages of cooling rectal temperature falls far more slowly than esophageal temperature. When the temperature has dropped to 90 to 91° F. by esophageal recording, the patient is removed from the ice bath to the operating table, and surgery is begun. The temperature will drift downward another 5 degrees. If the temperature falls below 85°, water warmed to 110° F. is run through a water mattress beneath the patient. Our experience indicates that temperatures below approximately 85° F. invite heart block, excessive bradycardia and fibrillation.

After exposing the heart through a transsternal incision at the base of the fourth intercostal space, the interior of the right atrium is explored with a finger through an incision in the auricular wall to confirm the diagnosis of a secundum defect and to determine its site and size. The rather laborious transsternal incision rather than a longitudinal incision is necessary to provide adequate exposure of the hilus of the left lung.

The superior and inferior venae cavae, the main pulmonary artery, the aorta and the hilus of each lung are isolated and surrounded with umbilical tapes. With a long curved, fine toothed forceps, a lip of the atrium is occluded at the proper position as determined by the previous finger exploration. An incision is made in this clamped-off segment, and holding sutures are placed on each side of the cut and at both its ends.

When all is in readiness, the cavae are occluded for thirty seconds to allow the heart completely to empty itself. Then the aorta and pulmonary artery are clamped, and the hilus of each lung is occluded by drawing up the tapes. To prevent blood from flowing back from the lungs into the left atrium and obscuring the field, it is important to occlude both hili. Physostigmine, about 0.5 cc. of a 1:4000 solution for a four-year-old child, is injected into the aorta proximal to the clamp to slow heart action.

The clamp is removed from the atrium, which is rapidly cleared of blood. To lessen the chances of air entering the left atrium, it is important not to push the aspirator through the defect. The defect or defects are quickly closed with running sutures of 000 silk. The atrium is allowed to fill with blood, and the incision in it is occluded by reapplying the toothed clamp. The clamps are promptly removed from the aorta and pulmonary artery, and the ligatures about the hili are released. The cavae are opened slowly to prevent sudden overfilling of the somewhat hypoxic heart. As soon as the heart is beating normally the opening in the right atrium is closed with running silk sutures. Although it is said that under hypothermia it is safe to stop circulation for eight minutes, the spectre of damage to the patient's brain by hypoxia


Aorta
Pulmon art
SVC
Left hilus
Right hilus
Rt. auricle
IVC
Interatrial defect

makes it desirable to try to limit interruption of circulation to not more than five minutes In one instance the circulation was stopped for seven minutes without sequelae of any kind

RESULTS Ostium secundum defects have been closed under direct vision in twenty-two patients under hypothermia without mortality or sequelae due to interrupted circulation Ventricular fibrillation occurred a number of times immediately after operation but was promptly corrected by electric shock In two instances fibrillation was due to a few bubbles of air in a coronary artery By strenuous massage the air was forced out of the vessel and a normal beat re-established

Eight ostium primum defects were closed with the aid of extracorporeal circulation as described in the chapter on Interventricular Septal Defects There was no mortality in this small group of cases Two attempts were made to close atrioventricular communis defects Both children died The associated abnormalities of the tricuspid and mitral valves make adequate closure of these defects next to impossible

General Considerations

Before hypothermia and extracorporeal circulation were well established, we operated upon twenty-four patients with a diagnosis of interatrial septal defect With no guide but a finger in the atrium of the beating heart we attempted blind closure of the defects utilizing various methods of suture We had lots of trouble A few defects were inadequately closed, two patients died after operation, and a third child, seven years old, with an ostium primum defect and a huge heart died after exploration only, Kathy was her name About six weeks after her death I received a check from her father for $7 16 and with it a letter which stated "Unprompted, Kathy's friends in the neighborhood collected this money and want you to have it to help some other little girl" Stimuli to renewed efforts come from unexpected sources

The uncertainty of being able adequately to close an uncomplicated interatrial defect in the time allowed under hypothermia and the chances of encountering defects and difficulties not anticipated argue for the routine use of extracorporeal circulation Against its use are the dangers of failure inherent in machines and the tremendously increased cost to the patient

We shall for the moment continue to use hypothermia for direct-vision closure of the uncomplicated relatively small interatrial defect of the secundum type, so typical that it cannot be mistaken, but shall use extracorporeal circulation for closure of all others However, from the trend of present day experience reported by many workers it appears that the heart-lung machine will replace total body hypothermia within the next few years for closure of all atrial septal defects

Interventricular Septal Defects

The opening statement made in the chapter on Interatrial Septal Defects is equally applicable here

General Observations

Interventricular septal defect occurring as a single lesion and in conjunction with infundibular stenosis is the most common congenital heart lesion and from the standpoint of management, probably the least well understood

The defects vary in size from a few millimeters to 15 mm in diameter in most cases to occasional complete absence of the interventricular wall. They occur most commonly in the membranous portion of the septum confluent with the origin of the aorta.

The two most important factors which determine the clinical course of a patient with an interventricular communication are the size of the defect and the associated or consequent pulmonary hypertension Why interatrial defects are less commonly complicated by pulmonary hypertension than interventricular defects is not clear It has been suggested that the transmitted impulse of the more powerful left ventricle to the lungs is a factor

Clinical and laboratory findings indicative of an interventricular defect are fairly typical, but vary tremendously in degree, depending upon the size of the defect The child with a small defect (Roger's disease) has normal color, is well, and has no exercise intolerance Only the heart murmur is of concern to the parents The murmur is harsh, extends through systole and is best heard near the fourth interspace to the left of the sternum A thrill if palpable, is slight The heart is quiet, and the apex beat is accompanied by no thrust against the chest wall The pulmonary second sound is unaccentuated In the roentgenogram a normal cardiac silhouette is seen Electrocardiographic tracings manifest no sign of heart strain Few of these patients are subjected to cardiac catheterization, because absence of symptoms does not warrant the procedure

The child with a large defect also may be well and surprisingly free from symptoms in view of the physical and laboratory findings A harsh, raucous systolic murmur is heard A thrill is practically always palpable over the lower sternum and is of considerable diagnostic significance A hyperdynamic heart beat at the apex is indicative of a large left-to-right shunt, a heaving impulse at the lower left sternal border suggests advanced pulmonary hypertension The pulmonic sound is mildly or acutely accentuated, depending upon the degree of pulmonary resistance In roentgenograms enlargement of the heart and an increased vascularity of the lung fields are seen Electrocardiograms often show both left and right ventricular hypertrophy, if pulmonary tension is high, right ventricular hypertrophy predominates

Cardiac catheterization studies are of primary importance for determining the volume of left-to-right shunt in the ventricles and for measuring the degree of vascular resistance in the pulmonary circuit All children with sizable defects have some elevation of right ventricular pressure The size of the shunt, i e , the volume of blood flow from left to right, is significant, but far less important than right ventricular pressure A large flow may be associated with low pulmonary pressure and a small flow with high pressure The latter condition is of much more serious prognostic import from a surgical standpoint

Such are the basic and elemental findings characteristic of the average interventricular septal defect Variations from the typical require diagnostic acumen of the pediatric cardiologist In fact, the average cardiac surgeon will have a bit of trouble in making an unchallengeable

diagnosis even in classic cases and will not be content to advise operation on the basis of his own findings

Indications for Operation

Repeated appraisal of the child's condition discriminating attention to heart murmurs and thrills skilled interpretation of changes in successive electrocardiograms and experienced evaluation of cardiac catheterization data—all these are essential before it is decided to advise operative closure of a defect. Between those patients who probably never will require operation and those who because of advanced disease are no longer eligible for operation lies a large group who may be benefited by closure of the defect While surgeons are directing their energies towards perfecting a heart lung machine cardiologists are striving for more clear-cut indications for operation

Children with interventricular septal defects amenable to surgery may be divided into three groups

1 Infants with large defects and early heart failure are at present a problem to which no answer has yet been written. These infants under weight, below par subject to frequent severe respiratory infections have large hearts are frequently in and out of congestive failure and look as though they were going to die Some do others rather miraculously at age one to two years improve Their hearts actually get smaller and they thrive gain weight, learn to walk and in general appear normal. Whether these infants improve because pulmonary vascular resistance lessens the volume of blood flow from left to right or because the size of the defect in the ventricular wall remains stationary while the heart grows and thus indirectly decreases the shunt is a matter of dispute. The former opinion seems more tenable Neither may be correct

Before indications for or against early operation can be written in ink these questions need to be answered Shall operative closure of the defect be advised for the desperately ill infant, or shall operation be delayed until the child has improved? While awaiting improvement is the golden opportunity of preventing irreversible pulmonary hypertension lost? Which is worse high operative mortality in infants near congestive failure or later high mortality from pulmonary hypertension? Our attitude at present is to await spontaneous improvement before advising operation

2 In group two are the patients who are welcomed by the surgeon They are the children who have moderate sized defects few general symptoms some enlargement of the heart, minimal electrocardiographic evidence of right and left heart strain a large left to-right shunt and above all right ventricular pressures *below 80 mm of mercury*

3 In group three are placed those children who are beginning to

show signs of weakening, who have large hearts, vascular lung fields, diminishing left-to-right shunts, dominant right heart strain and, most important, pulmonary pressures *above 80 mm of mercury* When right ventricular pressures reach or exceed systemic pressures, the outlook is bleak with operation and rather hopeless without The child who is cyanotic at rest and more so after exercise is an exceedingly poor surgical risk because of irreversible pulmonary vascular changes This last statement about irreversible changes may be wrong Too few patients with severe pulmonary hypertension have survived operation and too little time has elapsed to evaluate survivors to know what the course of pulmonary vascular sclerosis will be It is known that the child with a patent ductus arteriosus complicated by pulmonary pressure above systemic pressure usually dies after operative closure of the ductus

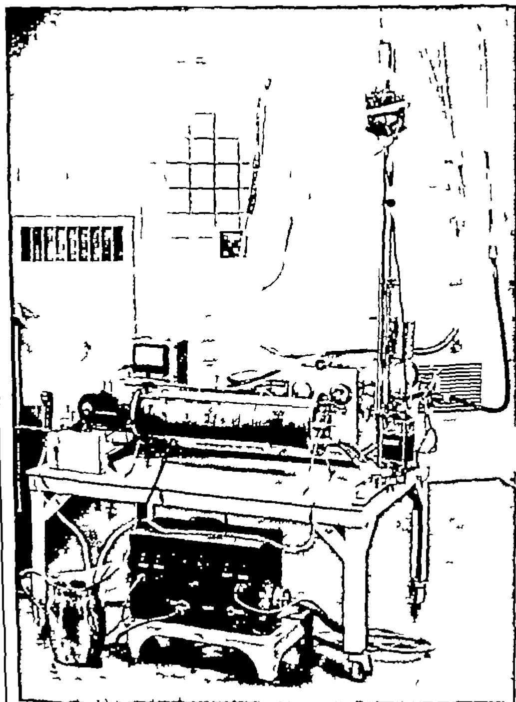
Surgical Treatment

Sporadic and ineffective attempts at blind closure of interventricular defects have given way to direct closure through the opened ventricle while respiration and circulation are maintained by a heart-lung machine To Lillehei and the group at the University of Minnesota goes the credit for opening this difficult but fascinating field of cardiac surgery Gibbon was the first to construct a workable heart-lung apparatus

Pumps to replace temporarily the function of the heart are readily available, but oxygenators which will physiologically perform the function of respiration are still in the process of development Bubble, membrane and disc oxygenators are now being used in various centers The modified Gibbon heart-lung machine is a complicated, beautifully functioning apparatus equipped with many safety devices and control gadgets The only barriers to its general use are the original cost and the upkeep

We spent three years experimenting with bubble and membrane oxygenators, never could make them work satisfactorily, and finally discarded both in favor of the disc oxygenator popularized by Kay and Cross The idea of a disc oxygenator was originally suggested by Bjork and some other ingenious Swedes Our basic apparatus for extracorporeal circulation now consists of a Sigmamotor pump, Kay-Cross oxygenators of varying sizes and the blood-collecting chamber and filtering system perfected by Gross

Clinical application to seventy-five patients during the past year in which extracorporeal circulation has been in use at the Children's Memorial Hospital has proved the apparatus satisfactory The oxygenator is effective, nontraumatic to blood, easy to assemble and simple enough in construction to be relatively free from mechanical failure We have lost no patient because of failure of this heart-lung machine



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Patent Ductus Arteriosus

Twenty years ago a young man just out of his surgical residency became convinced that a patent ductus arteriosus could be closed surgically His name was Robert E Gross He studied every angle of attack upon this congenital anomaly and proceeded methodically to familiarize himself with the anatomy of the large vessels at the base of the heart and to perfect on animals the technique of handling large, pulsating vessels On August 26 1938 he accomplished the first successful closure of a patent ductus arteriosus on a girl seven years old It seems absolutely incredible that only twenty years have passed since the door to surgical attack upon congenital cardiovascular anomalies was opened by an operation which today is so simple and common

Normally the ductus arteriosus closes shortly after birth, having fulfilled its fetal function of shunting blood from the pulmonary artery

Let this be made crystal clear The entire procedure of extracorporeal circulation from admission of the patient to the hospital to his discharge is tremendously exacting and time-consuming Much equipment is needed, services of many people—house staff, nurses, technicians—are essential, and devoted attention to innumerable details is mandatory Surgeons contemplating open heart surgery cannot master the total technique by reading books, looking at illustrations or even watching others work Such guides are valuable, but long practice in the animal laboratory—that is the place to make the inevitable mistakes—is a prerequisite to clinical application

It might be well to add that open heart surgery is best done by young men whose fingers are nimble, whose minds are agile and whose coronaries are pliable Lest this statement be misunderstood, I hasten to explain that my associates, Doctors Riker, DeBoer and Baffes, operate upon the difficult cases while I now stand by and cheer.

RESULTS. Fifty-eight interventricular septal defects have been closed—forty-three isolated defects and fifteen associated with some degree of infundibular stenosis No mortality occurred in 37 patients with simple defects whose pulmonary pressures were below 80 mm of mercury Five of 6 patients whose pulmonary pressure was above 80 mm of mercury died after operation

Pulmonary hypertension indubitably is the number one problem in patients with interventricular septal defects, and its solution probably will be found in better medical therapy or earlier operation

been described by a farm boy "like feeling bees in a bag." This thrill is more acutely felt by the hypothenar surface of the hand than by the finger tips

Bounding femoral pulses signify high pulse pressure A capillary pulse may be seen in the fingernails or in the turned-out lower lip compressed with a glass slide

Fluoroscopic examination to demonstrate hilar dance in the lungs is often omitted because increased hilar markings are easily recognizable in the roentgenograms Minimal exposure to x ray irradiation is desir able Some degree of cardiac enlargement is usually demonstrable in the x ray films and a convex rather than concave shadow is seen in the region of the pulmonary segment

Electrocardiographic changes are usually minimal

A child with a typical patent ductus arteriosus is never blue is healthy and active In fact, parents so commonly remark that the child is more active than their other children that one can t but wonder whether double oxygenation of part of the blood acts as a supercharger

About ten years ago before the importance of pulmonary hyper tension as a complication of patent ductus was fully appreciated such a well known cardiologist as the late Stanley Gibson refused to make a diagnosis in the absence of a diastolic murmur He lived just long enough to call attention to his error A child with considerable pulmon ary hypertension does not have a diastolic murmur because pressure in the pulmonary vessels is as high as systemic pressure during diastole A ringing pulmonic second sound alerts one to the possibility of pul monary hypertension A tinge of cyanosis in the lips or fingers during exercise suggests that pulmonary hypertension is severe enough to reverse the blood flow from right to left Continuous cyanosis even though mild is an ominous sign of excessive pulmonary hypertension.

An unusual case came under our observation a ten-year-old child with cardiac enlargement no murmur prominence of the pulmonary segment, clouded lung fields and a snapping pulmonic second sound. The history of having had a murmur during early childhood suggested the possibility of a patent ductus with pulmonary hypertension. Angiograms confirmed such a diagnosis

Treatment

Cardiologists are now well agreed that all typical patent ductus (the plural is the same as the singular—fourth declension) should be corrected surgically during early childhood The operation is far simpler on a two-year-old child than on a teen ager and far simpler and safer than on an adult.

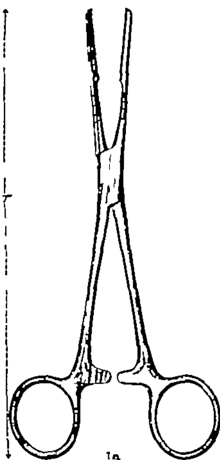
In the early years of operative treatment the ductus was occluded

to the aorta. Why the occasional ductus—about one in 2000 births—fails to close is unknown. Because a ductus not uncommonly closes spontaneously during the first month or two of life it is wise to be cautious in the interpretation of murmurs heard during the neonatal period. Misunderstanding between physicians may easily arise over the disputed absence or presence of a murmur in a baby. One physician hears it without a doubt and so informs the parents. The mother in a frenzy takes the child to another physician who hears no murmur. It is up to the second examiner for the sake of good public relations to explain that the ductus probably closed between visits and that both doctors were right.

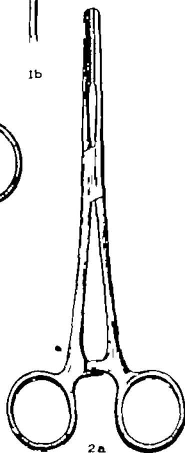
Diagnosis

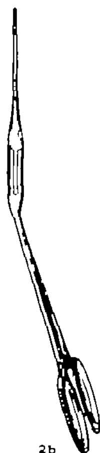
It is well to bear in mind that a ductus may remain open during infancy and be the cause of unexplained heart failure. The diagnosis is not easily made because the diastolic phase of the murmur is usually absent. A brisk systolic murmur in the characteristic location plus enlargement of the heart and no cyanosis should rouse suspicion of a patent ductus. Retrograde angiography is often necessary to clinch the diagnosis. A child six weeks of age was admitted to the hospital in unexplained heart failure. Edema was generalized, the liver was down to the crest of the ilium, a systolic murmur was heard at the second left interspace, and the heart was so enlarged that its left side lay against the lateral chest wall. Digitalis therapy brought about some improvement. With considerable trepidation a retrograde angiogram was made and showed a huge left-to-right shunt. At operation, performed as an emergency, a ductus larger than the aorta was found and successfully closed. Clinical improvement following operation in such cases is gratifying.

Diagnosis of a typical and uncomplicated patent ductus in children can be made almost without error on the basis of the murmur alone. At the second or third interspace to the left of the sternum one hears a rough, rumbling systolic murmur which goes well into and often all the way through diastole. After having been heard a few times it is practically unmistakable. The mother at times will say she can hear the noise when the child is lying next to her in bed. The size of the ductus is in a measure proportional to the intensity of the murmur. The less noisy murmurs are often heard only over a small area in the region of the pulmonary segment and cannot be picked up at the apex. In fact, in hurried examinations such as are conducted upon long rows of school children, in which the stethoscope is momentarily clapped on the chest over the region of the apex, the murmur can easily be missed. At the point of maximum intensity of the murmur a thrill can be felt, it has


1a


1b


2a


2b


3

by a single, snugly tied silk ligature It was soon learned that "ligation in continuity" led to recanalization For a time multiple ligatures were used, but still recurrences appeared Then Gross advised clamping, cutting and suturing to eliminate all chances of reopening Some surgeons still use multiple ligation plus a suture ligature through the middle of the ductus We believe that division and suture are not only preferable, but also safer The first twenty-one ductus operated upon at our hospital were closed with multiple ligatures and, although to the best of our knowledge none have recurred, I was unhappy with ligatures because of a near catastrophe In my eagerness to close a large ductus completely the ligature was drawn up snugly That last little tug was too much, and the ligature cut through the vessel wall Although the hole was repaired and recovery followed, the sight of blood cascading into the chest stimulated the search for suitable and safe ductus clamps

In 1947, quite by chance, the idea occurred to me of making a clamp with tiny teeth in the opposing jaws The teeth would have to be so tiny that they would grasp but not penetrate the vessel, so sharp that they would easily embed themselves in the outer wall of the vessel to prevent slipping, and so numerous as to distribute pressure equally to the entire vessel wall The hub of the clamp would have to be so built that the jaws could appose but not crush These ideas were presented to Mr Bruno Richter, an imaginative and capable instrument maker, who skillfully fashioned such clamps These original clamps with many modifications in size and shape are widely used today in all cardiovascular surgery As a matter of fact, the principle embodied in these clamps—I say this in a moment of lost modesty—has made some vascular operations possible and many safer

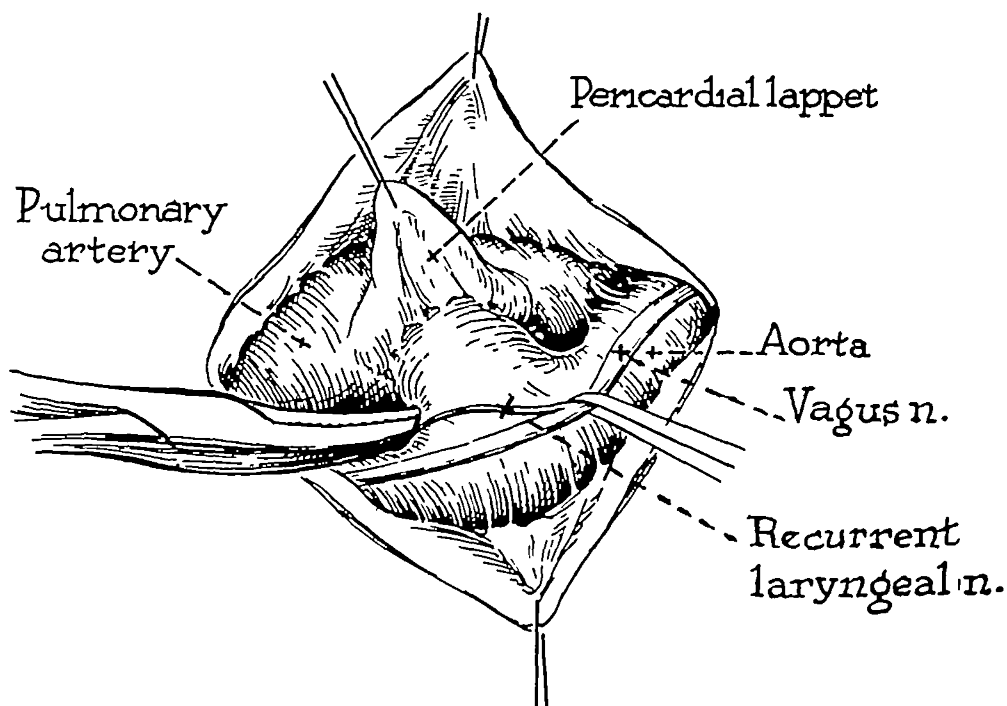
The operation is performed under ether-cyclopropane anesthesia With the patient on his right side the chest is entered through the left fourth interspace In children up to sixteen years of age a rib is never resected for this operation The vagus nerve and its recurrent laryngeal branch are promptly identified and serve as an infallible guide to the ductus The most important step of this operation is painstaking and thorough dissection of the ductus from surrounding structures A line of cleavage can be recognized between the adventitial wall of the ductus and surrounding tissue The dissection must be made through this delicate cobweb-like tissue Two steps are of the utmost importance The pericardial lappet must be dissected well off the ductus, and the fibrous tissue posterior to the ductus overlying the bronchus must be identified and dissected away from the ductus at both its aortic and pulmonary ends Unless these two simple maneuvers are assiduously attended to, the teeth of the clamps, when applied, will embed themselves in this tough tissue and allow the elastic stump of the ductus to roll out between the jaws of the clamp This misfortune has happened to us and to others

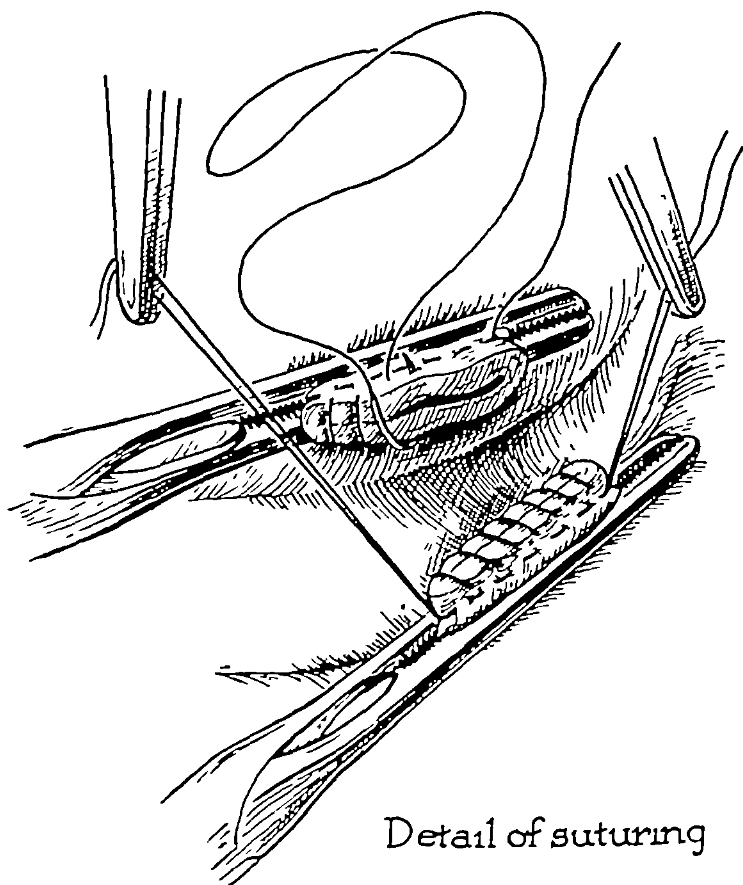
who have heatedly complained that the clamps were at fault. The diffi culty was due only to inadequate dissection

When dissection is complete two toothed clamps are applied one on the aortic end, the other on the pulmonary end of the ductus With a long thin bladed scissors the ductus is cut and each end sewed back and forth with a continuous suture of 5-0 Deknatel silk on a curved needle We routinely divide and suture all ductus The remainder of the operation and the postoperative care are standard and routine

A real problem presents itself in the child who has a ductus with pulmonary hypertension Although the diagnosis of patent ductus with pulmonary hypertension has been made before operation, we have advised operation on all these patients even though heart catheterization studies had demonstrated that pressure in the pulmonary artery was equal to or above systemic pressure After the ductus pulmonary artery and aorta have been exposed pressure readings in the pulmonary artery are recorded Then the ductus is clamped. If pressure in the pulmonary artery falls even so little as 10 mm. of mercury we know from experience that we may safely proceed with division and suture We have had five cases in which pulmonary pressure rose after the ductus had been clamped. We debated long whether to close or not to close the ductus Lacking knowledge about what to do and knowing that pulmonary hy pertension, if not remedied, would surely prove fatal we divided the ductus It appears now that pulmonary hypertension which exceeds sys temic pressure is not remediable One patient died on the operating table —our only operative death. Two children later died suddenly one six months and one two years after operation The other two are alive but sickly constantly on digitalis because of recurrent cardiac decompensa tion. It appears at this time that nothing can be done for excessive pul monary hypertension and that division of the ductus is of no avail Interestingly enough, those patients in whom pressure in the pulmonary artery fell even slightly when the ductus was clamped have gotten along well since operation.

No operation in the field of cardiovascular disease, congenital or acquired is so gratifying to surgeon and parents as division and suture of a patent ductus arteriosus After a successful operation one may tell the anxious parents that the difficulty has been remedied and that their child may look forward to full life expectancy Low operative mortality justifies the advice that all uncomplicated ductus should be operated upon at a convenient time after the diagnosis has been unquestionably made It has been our good fortune to have only one operative death referred to above in over 500 operations for patent ductus arteriosus performed by the attending and resident staff at the Children s Memorial Hospital during the past twelve years.





Detail of suturing

Coarctation of the Aorta

After the door to surgical correction of congenital heart defects had been opened by Gross in 1938 when he successfully occluded a patent ductus arteriosus, it is not surprising that coarctation of the aorta was tackled next. Almost simultaneously in 1945 Crafoord and Gross, working independently presented a practical method for resection of a congenitally constricted segment of the descending arch of the aorta. Except to the recorder of history it makes little difference who was first to describe this operation important only is the fact that the operation is sound and universally applicable

Adult type coarctation causes few symptoms in children below ten or twelve years of age Only after leading questions do they recall that their legs at times felt weak or cold or "went to sleep" Complaints of headaches, dizziness and other signs of hypertension are rare before fifteen years of age

Absence of femoral pulses in children is practically diagnostic of coarctation. Palpable pulse beats of intercostal arteries over the back confirm the diagnosis Notching of the under surface of ribs is rarely seen in roentgenograms before age seven years Retrograde angiograms are unnecessary to make a diagnosis in typical cases the hazard of an angiogram, although slight does not justify the procedure

It is wise to take the blood pressure in both arms If pressure in the left arm is 30 to 40 mm of mercury less than that in the right arm the surgeon should make preparations for unexpected variations in the operative procedure

Indications for Operation

The most desirable age for surgical resection of the adult type coarctation is between seven and twelve years Operation during infancy is apt to be followed by stricture at the suture line operation during the late teens and especially during the twenties and thirties is technically difficult and attended by more operative and postoperative complications

Operation is routinely advised for the child who has a persistently high systolic blood pressure in the arms and low pressure in the legs Observation for a year or more is appropriate for the child whose femoral pulses are weak, whose collateral pulses over the back are barely palpable and whose blood pressure in the arms and legs is near equal. The chances are, however that operation eventually will be necessary

It is difficult for parents to accept advice for a rather major operation upon their child who is in apparently excellent health. It is explained that the operation is a prophylactic procedure against later catastrophes and that it is least dangerous if done before irremediable changes in the heart and vessels have occurred.

Unanimity of opinion has not been reached by cardiologists and surgeons about what to advise for the infant who has coarctation of the aorta If the condition is an incidental finding and the child has no symptoms it is without question that operation should be delayed until the chosen age has been reached A nine month-old baby boy was examined for a heart murmur The femoral pulses were absent and the systolic blood pressure was 220 mm. of mercury The child remained well and grew normally although his blood pressure consistently ranged

Pathology

It is unfortunate that the word "coarctation" (from the Latin *coarctere,* to contract) has crept into medical literature Wedded to this awkward term by the common law of usage, we shall have difficulty discarding it for the more attractive stenosis or constriction

By far the most common site of coarctation of the aorta is in the immediate vicinity of the ductus arteriosus, and in medical parlance it is referred to as the adult type Although the term "adult type coarctation" is also an improper term, common usage likewise has forced its acceptance The constriction is usually abrupt In a few cases narrowing of the lumen of the aorta above and below the coarctation produces an hourglass configuration The constriction may and usually does appear at or near the origin of the ductus arteriosus It seems reasonable to suppose, although experimental proof is lacking, that the inborn elements which make a ductus close have spilled over into the wall of the aorta, where they fulfill their function and cause constriction

It is always amazing after removal of a coarctation to see how small the lumen of the resected segment is—usually not more than a few millimeters in diameter The variable number of thin-walled, dilated intercostal arteries which enter the aorta distal to the obstruction provide for collateral circulation

Infantile coarctation, also called preductal coarctation, has recently been more accurately labelled hypoplasia of the aortic tract complex by Dr Maurice Lev This term is appropriate because not only the arch of the aorta, but also the left ventricle itself, may be underdeveloped Varying degrees of hypoplasia may occur from a practically symptomless narrowing of the arch to complete atresia of the aorta and the left ventricle Obviously, the more severe and extensive the constriction, the more serious the symptoms Children having hypoplasia of the entire outflow tract die at or shortly after birth Moderate constriction of the arch is usually associated with other intracardiac anomalies

Diagnosis

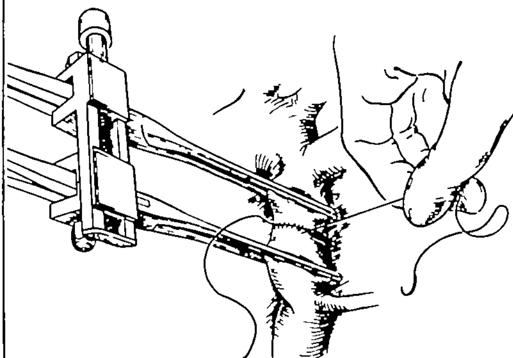
The frequency with which coarctation of the aorta is found is directly proportional to the persistence with which the examiner feels for femoral pulses A ten-year-old boy was admitted to the hospital with a diagnosis of acute appendicitis A resident who had just completed his service in cardiology examined the patient, then slid his fingers over the femoral artery in the groin and felt no pulse He found a systolic blood pressure of 220 mm of mercury in the arms and no measurable pressure in the legs After the boy's recovery from an appendectomy the coarctation of the aorta was resected

Umbilical tape

Ligated intercostal a.

trans-
fixed
duct.
art.

Excised portion of aorta



between 175 and 220 mm When the coarctation was resected at age six and one-half years, there still was minimal enlargement of the heart.

The infant with adult type coarctation complicated by heart failure presents a problem Some advise early operation We are inclined to carry the child on digitalis and, if possible, postpone operation until the suitable age Many of these children slowly creep out of heart failure, most likely because adequate collateral circulation develops and thereby relieves strain on the heart.

Operation

The patient is placed on his right side, and a curved incision is made beneath the left scapula To control bleeding from the large collateral vessels, I have found it helpful to insert my finger beneath the muscle and push upwards while cutting As the vessels come into view they are doubly clamped and cut Bleeding from those vessels which cannot be seen before they are cut is lessened by upward pressure with the finger It is essential to resect the fourth rib subperiosteally to provide adequate exposure

The aorta is dissected from surrounding structures entirely by sharp dissection with a scissors Umbilical tapes are placed around the aorta above and below the constriction to control bleeding in case of emergency and to assist in further dissection The ductus arteriosus, even though it appears to be closed, is isolated, doubly clamped and cut

Actually, the most dangerous step in resection of a coarctation is that of freeing the collateral vessels entering the distal segment of the aorta These large, thin-walled intercostal vessels are exceedingly fragile and rupture at a touch, and when they bleed, they bleed "like mad" A torn intercostal is hard to catch because the proximal end retracts between the ribs where it can't be seen A couple of experiences with these satanic bleeders makes a Christian out of any surgeon Only the intercostal vessels entering the upper end of the distal segment need be divided and cut to make room for the clamps

After the aorta has been well freed two toothed coarctation forceps are applied, and the constriction plus the narrowed portion of the aorta is excised We aim to cut away enough constricted aorta so that the final anastomotic site is of the same caliber as the aorta directly proximal to the anastomosis Our experience with this operation is limited to children sixteen years or younger, and that is very likely the reason we have always been able to coapt the ends and have never been forced to use a graft Extensive dissection of the arch of the aorta provides extra length in those instances in which a long narrow segment of aorta needs to be resected

If considerable traction is necessary to bring the ends of the aorta

Pure Pulmonary Stenosis

The proper name for this condition is congenital pulmonary valvular stenosis with intact interventricular septum but the universal tendency towards simplification has popularized the term "pure pulmonary stenosis" Although the important feature of this condition is stenosis of the pulmonary valve the word "pure" is a misnomer because there is almost always an associated defect of the atrial septum. Obstruction in the pulmonary valve varies all the way from a pinpoint opening in the center of the fused cusps to minimal contracture at the base of the valve In severe stenosis the fused cusps of the valve project into the pulmonary artery and resemble a truncated cone A defect of the atrial septum, usually an enlarged foramen ovale serves as an escape mechanism for blood the right ventricle is unable to force through the obstructed valve

together, a vise is applied to the clamps, and with a turn screw the ends are drawn into apposition and mechanically held while the anastomosis is performed Residents like the vise because it relieves them of the muscle-paralyzing task of holding the clamps and the fear of reprimand in case their hands should weaken at a critical moment

The ends of the aorta are anatomically apposed with a continuous suture of 5-0 Deknatel silk on a swaged-on needle The suture is interrupted once during the anastomosis The chest is always drained

RESULTS Resection of a coarctation of the aorta is a satisfactory operation No instance of postoperative bleeding from the anastomosis has plagued us—to date Immediate and late results are good, and operative mortality is low One death in our series of ninety-six resections of adult type coarctations occurred during operation upon a fourteen-month-old child with cardiomegaly due, we thought, to the constriction At post-mortem examination extensive fibroelastosis was found

A two-year-old child with cyanosis of the left arm, trunk and legs was explored and found to have stenosis of the aorta between the origin of the left common carotid artery and the left subclavian artery Blood flow to the distal aorta and the left subclavian artery from a widely patent ductus arteriosus accounted for the demarcated cyanosis The ductus was cut and its distal end sutured, not without difficulty, to the arch of the aorta Death due to right-sided heart failure occurred a few hours after operation To date no effective operation has been devised for this complicated anomaly

Treatment

Valvulotomy is the only effective treatment. Brock, of England, popularized a method of introducing a rigid, diamond shaped valvulotome into the right ventricle and thrusting it through the stenotic valve. After cutting the valve, graduated dilators were inserted until pulmonary flow was unimpeded

To lessen trauma to the ventricular wall and to decrease blood loss we devised a valvulotome with retractable blades and a similarly func tioning dilator which could be widely expanded after introduction into the constricted area Besides these instruments we also had made a tiny valvulotome for quick use in very cyanotic infants with tremendously enlarged hearts The small valvulotome consists of a cataract knife blade soldered to a long small-caliber rod attached to a regular scalpel handle The operating room nurses to distinguish it from the regular instruments refer to it as the "toad stabber"

During recent years valvulotomy under direct vision has become popular and in most patients is the treatment of choice Some perform this operation under hypothermia others prefer to do it with the aid of extracorporeal circulation.

Our method of surgical treatment at present varies somewhat depending upon the patient s age and the severity of symptoms Rapid and simple operation is essential in an infant a few months old, deeply cyanotic, with severe cardiomegaly in or near heart failure The chest is entered by a left submammary incision through the fourth interspace The second, third and fourth ribs are cut at the sternal junction. The pericardium is opened and the diagnosis quickly confirmed by feeling a jet of blood spurting through the stenotic valve and by observing the dilated pulmonary artery A purse-string suture is placed in the wall of the right ventricle, and, if heart action is poor the tiny valvulotome ("toad stabber") is quickly thrust into the ventricle and the stenotic valve cut. It is amazing to see the child s color suddenly change from blue to pink. Cardiac muscle responds to increased oxygenation with a quick ened and more forceful beat. If it is apparent that the infant s condition will tolerate further surgery the pulmonary constriction is widely opened with the regular valvulotome and dilator

For eight years and approximately 100 patients we used the transventricular approach and by the closed method cut and dilated the constricted pulmonary valve Our aim at operation was to reduce right ventricular pressure to approximately 250 mm of water Often the valvu lotome and dilator had to be introduced a number of times to accomplish this objective The operation is safe easy quick and immediately quite satisfactory Only one death occurred in this series The objection—a real one—to this procedure we slowly learned was the disheartening fact that stenosis recurs especially in older children

Diagnosis

Symptoms, obviously, are in direct proportion to the degree of cyanosis and consequently vary from minimal to incapacitating Patients are brought to the cardiologist because of cyanosis and/or disability or because an unsuspected heart murmur was found Cyanosis may be deep or barely discernible during violent exercise The red blood cell count is usually elevated, but, in general, is not as high as in patients with tetralogy of Fallot The murmur, confined to systole and heard over the pulmonic area, may be a mere scratch in an infant with severe stenosis or a turbulent roar with vibrating thrill in older children with moderate stenosis The second pulmonic sound is diminished or absent

Roentgenographically, the lung fields show diminished markings in the cyanotic and little variation from normal in the noncyanotic As is to be expected, heart size, varying from normal to minimal enlargement of the right ventricle, to enormous distention of both right atrium and ventricle, is a direct response to the degree of valvular obstruction The pulmonary artery just beyond the valve is usually considerably enlarged It is an interesting phenomenon that the pulmonary artery dilates when obstruction is in the valve itself and remains small when obstruction is in the infundibulum as in tetralogy of Fallot Holman explains this dilatation of the pulmonary artery scientifically the jet of blood spurting at high pressure through the constricted valve expends its energy in all directions, strikes the sides of the vessel and makes it expand It sounds reasonable

Electrocardiographic tracings show deviation of the axis to the right —a peaked P wave in lead II

Cardiac catheterization studies are usually in order, not to make a diagnosis except in atypical cases, but to determine the gradient of pressure between pulmonary artery and right ventricle Normally, pressure in the right ventricle varies between 20 and 30 mm of mercury In pure pulmonary stenosis it may go as high as 200 mm

Prompt operation is indicated in all patients with pure pulmonary stenosis who are cyanotic, or have any degree of disability, cardiac enlargement or a right ventricular pressure of 100 mm of mercury or more Occasionally one sees a patient with no symptoms, little if any cardiomegaly, but a ventricular pressure of approximately 100 mm It is generally agreed that such a patient should choose surgical correction as a prophylactic measure against cardiac overloading Ventricular pressure of approximately 75 mm in patients with no symptoms and no cardiac enlargement requires observation rather than operation Naturally, one doesn't catheterize such a person every six months Symptoms, heart size, electrocardiographic changes are reviewed from time to time, and when it appears indicated, a second catheterization is performed and the patient advised accordingly

Tape around rt. pulm. art

Left pulmon artery

Aorta

SVC

IVC

V. Samter

Detail of stenosed pulm valve

Except for emergency operations in infants as described above, we now, with the aid of hypothermia, open the pulmonary artery in a bloodless field and under direct vision completely release the stenotic valve cusps The child is anesthetized and packed in ice When the rectal temperature reaches 91° F, the patient is taken out of ice and placed on the operating table on a water mattress The temperature will drift to about 85° F If it continues downward, warm water is run through the mattress and the operation temporarily halted until the temperature returns to 85° F A Y-shaped incision is made in the skin and the sternum split longitudinally The superior and inferior venae cavae, the aorta and the right and left pulmonary arteries are isolated and surrounded by tapes Four holding sutures are placed in the wall of the pulmonary artery just distal to the valve to steady the vessel while a curved toothed clamp is applied longitudinally A ¾ to 1-inch long incision is made in the occluded lip of the pulmonary artery

The venae cavae are occluded, and after allowing thirty seconds for the heart to empty itself the aorta and pulmonary arteries are occluded For a six-year-old child 0 5 cc of 1 4000 Prostigmin is injected into the aorta proximal to the clamp to slow heart action The clamp is removed from the main pulmonary artery and the tip of the funnel-shaped obstructed valve grasped with a hemostat With a scissors the valve cusps are cut all the way to the base along their lines of fusion A finger is inserted through the pulmonary artery into the ventricle to feel for infundibular stenosis, which, if present, is removed with a rongeur forceps The clamp is reapplied to occlude the incision in the main pulmonary artery As it is being applied the ligatures on the pulmonary artery are released to allow blood to fill up the vessel and the ventricle The aortic clamp is removed, and the ligature on the superior vena cava is loosened slowly to prevent sudden overloading of the heart As soon as the heart takes over with a good firm beat the inferior vena cava is released The cut in the pulmonary artery is sutured with 5-0 Deknatel silk, and routine closure of the wound follows When the temperature reaches 94° to 95° F, the child is sent to the recovery room

The time of occlusion of blood flow through the heart is usually about two minutes by a stop watch in uncomplicated valvular stenosis and twice that time if there is an additional infundibular stenosis We do not like to stop circulation for more than an absolute maximum of five minutes—less if possible Although to date it has been unnecessary, we plan for that case in which the open portion of the operation cannot be completed in near four minutes to clamp the incision in the pulmonary artery, loosen all the tapes on the inflow and outflow tracts and restore normal circulation After the heart has been beating normally for about ten minutes circulation will again be interrupted to complete the intracardiac procedure Nothing in the world is worse than permanent brain damage as a result of too prolonged interruption of circulation

Tetralogy of Fallot

The first ray of light broke over the dark field of congenital cyanotic heart disease in 1945 when Blalock and Taussig demonstrated that cyanosis due to tetralogy of Fallot could be relieved surgically. Parents of "blue babies" were thrilled to learn that something could be done to relieve their helpless children of stifling oxygen want. Interest of physicians and cardiologists was quickened by the knowledge that this almost universally fatal deformity of the heart was at least in a measure remediable. Even junior medical students beat off afternoon lethargy to learn the fundamentals about this disease.

Pathology

Pathologically, Fallot's tetrad is a combination of stenosis in the outflow tract of the right ventricle, overriding of the aorta, a defect in the interventricular septum, and hypertrophy of the right ventricular wall. To the original and classic description have been added many

To permit less hurried surgery, some cardiac surgeons prefer to do all open valvulotomies with the aid of extracorporeal circulation. Certainly there can be no objection to this if sufficient personnel is available to draw and prepare blood and keep the extracorporeal machinery in faultless working order I may regret the following sentence by the time this book is published It hardly seems justifiable to spend so much effort and expense for an operation which can be done safely under hypothermia in two to four minutes Perhaps by good fortune we have escaped the pitfalls of hypothermia and interrupted circulation, such as ventricular fibrillation and brain damage To date, in forty-one patients operated upon for pulmonary stenosis under direct vision there has been no serious postoperative morbidity and no mortality

Results of closed valvulotomy, as stated above, have been good in infants and little children, presumably because the constricting tissue is soft and easily cut and torn In some older children, especially in or near teen age, the valvulotome presumably did not cut the valve completely, nor did the dilator tear the valve all the way to its base Immediate clinical results were excellent Children who had been completely helpless could engage in strenuous activities soon after operation Although the majority have maintained these good results, a few children have had recurrence of symptoms To date we have had to perform open valvulotomies on six children who previously had undergone transventricular valvulotomy We were surprised at the second operation to note how the edges of the valves had grown together Of course, time only will tell whether recurrences will also follow complete incision of the constricted valves under direct vision It is now our policy to do open valvulotomy under hypothermia in all but poor-risk infants

The first child upon whom we operated for pure pulmonary stenosis in 1950 was a 23-day-old infant boy with a tremendous heart and a pulsating liver near the iliac crest As the pericardium was opened the heart became very slow and weak The anesthesiologist said, "If you're going to do something, you better be quick about it" The small cataract knife blade was literally jabbed into the right ventricle, and with a few quick motions, keeping the sharp edge of the blade towards the lumen of the vessel, a number of radial incisions were made in the valve leaflets Immediately the rate and force of the heart beat improved Fearing that we might be tempting fate by using a dilator, we left well enough alone Evidently good fortune smiled upon us that day because the child made an excellent recovery A diastolic murmur has developed indicative of pulmonary regurgitation, but that seems to be no handicap A check-up examination after this chapter was written indicates that some recurrence of stenosis is developing It's hard at times to face the truth

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variations Infundibular obstruction may be tubular or diaphragm-like, and may be situated anywhere in the right ventricle from apex to pulmonary valve Combinations of infundibular and pulmonary valve stenosis are not uncommon Variations in size of the pulmonary artery extend all the way from atretic to normal, and in cases of valvular stenosis to great dilatation Overriding of the aorta over the interventricular defect varies from a minimal degree to as much as 90 per cent Hypertrophy of the right ventricle is not a pathologic finding, but rather the physiologic result of overwork against obstruction

Diagnosis

The symptoms and findings in a typical case of tetralogy of Fallot are so specific that any physician can make a diagnosis Cyanosis is the outstanding feature Often it does not manifest itself during the first few months of life because the ductus arteriosus remains open and shunts some blood to the lungs As the ductus closes, cyanosis deepens, and its degree is determined by the extent of pathology in the heart The red blood cell count ranges from slightly above normal to as high as 13 million cells per cubic centimeter The average is 7 million

The severely cyanotic infant, a year or two of age, is a pitiable creature—fussy, unhappy and hopelessly spoiled Crying is apt to bring on an attack of syncope, and so the parents don't let the child cry Very early in life the infant learns the power of its weapon and uses it effectively

A history of slow physical development is routinely obtained If the child is old enough to walk, there is almost always a history of squatting In fact, squatting, rather than sitting or lying down, to relieve dyspnea is so characteristic that it has been accepted as an important sign If there has been no history of squatting, suspicion arises as to whether the disease actually is tetralogy of Fallot

Clubbing of fingers and toes appears during late infancy and is conspicuous in older children A systolic murmur is heard over the second or third interspace just to the left of the sternum, and varies in intensity, depending upon the degree and type of stenosis A slight "medical murmur"—not audible to the surgeon—usually means a small pulmonary artery and almost complete occlusion of the outflow tract A rough, scratchy murmur is indicative of a large pulmonary artery and subvalvular stenosis The pulmonic second sound is weak If the murmur goes into diastole, the child does not have tetralogy of Fallot

Fluoroscopic and roentgenographic studies will show diminished markings in the lung fields and a normal-sized or boot-shaped heart Right axis shift will invariably be seen in the electrocardiogram For a diagnosis of typical tetralogy of Fallot nothing further is needed Atypical cases

require the utmost in diagnostic acumen and the help of angiocardiograms and catheterization studies

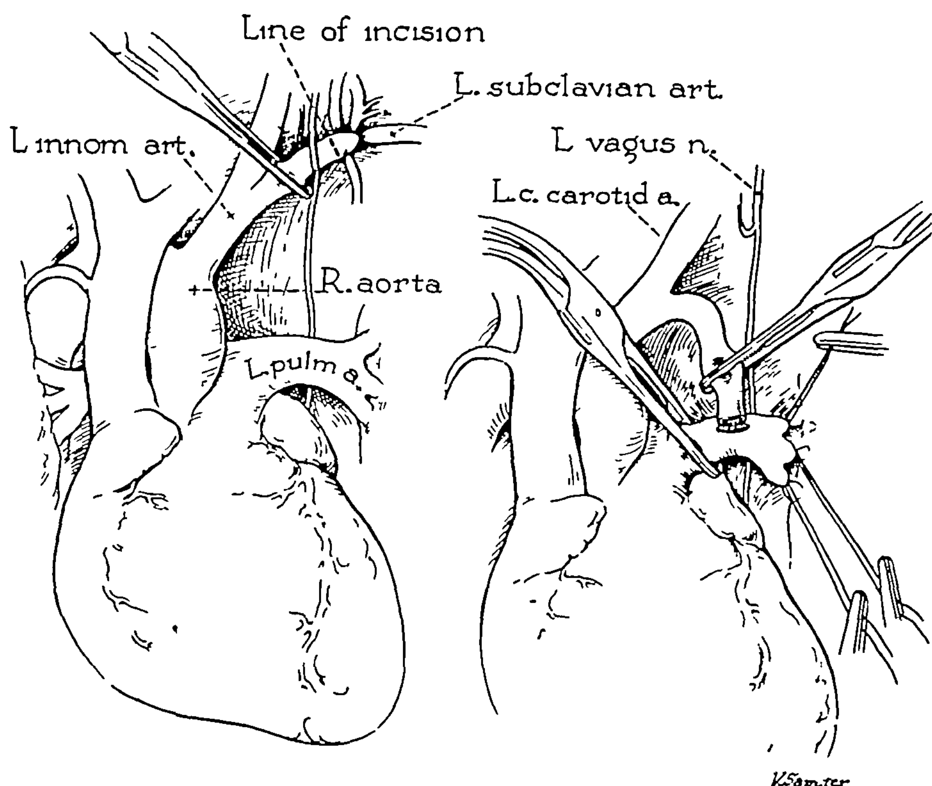
History of Surgical Treatment

The story of the development of the original surgical treatment for the relief of cyanosis due to the tetralogy of Fallot is an interesting example of serendipity For years Blalock had been working on the problem of arteriosclerosis Since sclerosis occurs in the aorta with high intraluminal pressure, but not in the pulmonary artery it appeared altogether logical that blood pressure might be an etiologic factor To raise the pressure in the pulmonary artery in animals Blalock anastomosed a systemic vessel—the subclavian artery—to the pulmonary artery Nothing happened to the pulmonary vessels but such an anastomosis was shown to be possible The successful anastomosis led to the idea suggested by Dr Helen Taussig "Why not do such an operation on a child with congenital pulmonary stenosis to increase the blood flow to the lungs?" Why not? The technique of subclavian pulmonary anastomosis was perfected and parents were found who were willing to have this operation attempted upon their children A number of successful operations opened the door to effective treatment of congenital cyanotic heart disease The story was told around the world.

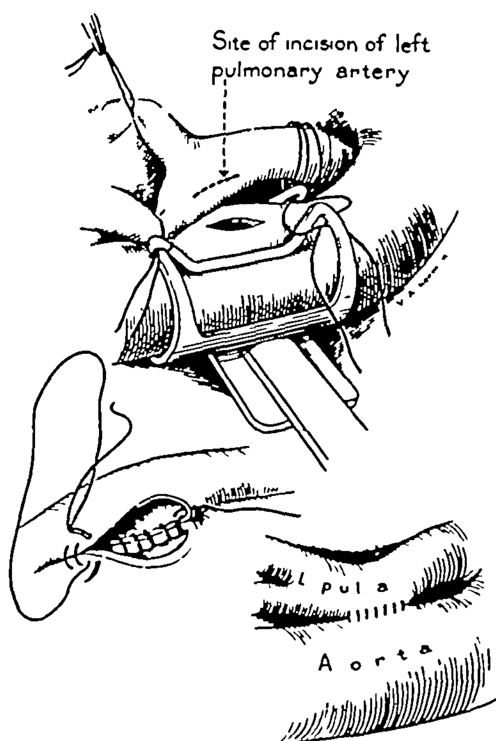
In 1946 I attended a clinicopathologic conference at the Children s Hospital in Boston where the organs of a child two years old were shown This child seemed perfectly normal until about six months of age, when cyanosis first appeared. At age eighteen months cyanosis was deep and attacks of syncope became frequent and finally proved fatal. It was suggested during the discussion that the patent ductus arteriosus probably accounted for the delayed onset of cyanosis—a fact well known today If a patent ductus could prevent cyanosis why not create an artificial patent ductus to relieve cyanosis?

Dr Sidney Smith and I worked out on dogs a method of performing aortic pulmonary anastomosis to accomplish this purpose This procedure was made possible by construction of an aortic clamp which partially occluded the aorta, and furnished a lip to which the pulmonary artery could be sewn Partial occlusion allowed enough blood to flow through the aorta to maintain adequate circulation while the anastomosis was being performed Longitudinal and parallel cuts 6 mm long were made in the aorta and pulmonary artery and a side-to-side anastomosis was performed. These 6-mm. incisions provided after anastomosis a shunting channel approximately 4 mm in diameter We reasoned that a shunt of such size would relieve cyanosis but not place too great a strain on the heart

In 1948 Brock of England devised a new attack upon the tetralogy



Blalock procedure:
anastomosis of subclavian-pulmonary arteries.



Potts-Smith procedure:
anastomosis of aorta-pulmonary artery

of Fallot aimed at removing the obstruction in the right ventricle At operation he opened the pericardium exposed the heart, and by palpation located the obstructing tissue in the right ventricle A purse-string suture was placed in the ventricular wall proximal to the obstruction Inside this suture used to lessen blood loss, an incision was made in the ventricle and through it with a rongeur forceps the obstructing infundibulum was removed bit by bit

Subclavian-pulmonary or aortic pulmonary anastomosis and infundibulectomy all have the disadvantage of being noncurative procedures. Nevertheless these operations and their variations do relieve cyanosis, are attended with relatively low mortality and do for a time improve the patient's clinical condition remarkably

From 1946 to 1956 immediate mortality following aortic pulmonary anastomosis remained the same approximately 8 per cent, in 600 patients operated upon at the Children's Memorial Hospital (During the past two years shunt operations have been almost exclusively confined to severely cyanotic infants requiring emergency relief) In children above three years of age the operative mortality remained at about 4 per cent in children below three years of age the mortality was 15 per cent. The reason for this discrepancy is not age, but degree of pathologic changes

Long term studies of patients after all types of shunt operations have shown similar results about 70 per cent are clinically tremendously improved approximately 15 per cent are somewhat improved, and 15 per cent die—during or immediately after operation, or in the course of a few years Statistics vary somewhat, depending upon methods of grouping and reporting.

Poor results are beginning to manifest themselves in children upon whom shunt operations were performed eight to ten years ago In recent years we have seen a few patients who have lost the diastolic phase of the murmur produced by the artificial ductus and have again become cyanotic. Catheterization studies confirm the disturbing fact that these children have pulmonary hypertension Why this complication should occur in some patients and not in others, although the operations were identical and the findings at operation similar we do not know The first aortic pulmonary anastomosis was performed in 1946 on a nineteen month-old child who was deeply cyanotic with a red blood cell count of 10,300 000 per cubic centimeter and who was completely helpless and subject to many attacks of syncope daily Now in 1958 she is thirteen years old healthy and active, in junior high school Her heart is only slightly enlarged. She is active—too active to suit her mother Last summer she pedalled fifteen miles on a bicycle in one day Why can't all results be as good?

It is appropriate to repeat that the shunt operations and infundib-

ulectomy are makeshift procedures which alleviate but do not correct intracardiac defects and offer little hope for normal life expectancy

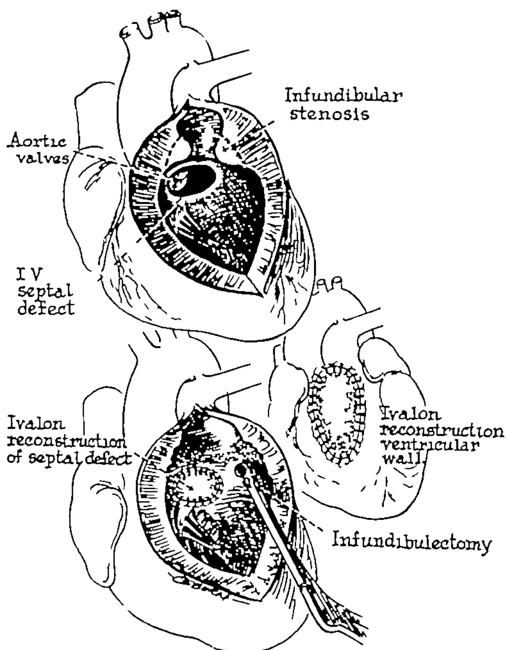
Present Surgical Treatment

Lillehei and co-workers originated and popularized intracardiac correction of pathology common to Fallot's tetrad There is little question that resection of the infundibular obstruction and closure of the interventricular septal defect is the ideal procedure The operation, attended by a rather startling mortality during the developmental period, is now being done in some cardiac centers with an average mortality rate of approximately 20 per cent, it is lower than this in clinics which report current mortality from time to time Occasionally it has appeared that an amateur player allowed himself a few "mulligans" on the intracardiac golf course It is rapidly becoming established that most tetralogies should have benefit of the definitive operation

At this time (1958) it is difficult to decide what type of operation should be advised for the severely cyanotic infant below six months or one year of age who has a minimal systolic murmur indicative of a tiny pulmonary artery, is subject to repeated attacks of syncope, and needs immediate surgical relief Open heart surgery on these tiny infants at present is attended with a high mortality rate At the moment we advise a shunt operation as an emergency lifesaving procedure with the understanding that further surgery may be necessary at a later date Intracardiac correction of the tetralogy of Fallot, being new, is beset with technical difficulties, unpredictable bleeding tendencies, mechanical inadequacies of artificial lungs and hearts, and with many postoperative complications However, it is safe to prophesy that within a few years these obstacles will have been overcome and that operative mortality will have been reduced to an acceptable and irreducible minimum Our personal experience with open heart surgery for tetralogies, although limited, is satisfactory Fifteen open heart operations have been performed on children with tetralogy of Fallot There have been three postoperative deaths

Long-term results following these operations are still fragmentary and inconclusive A number of questions remain to be answered Will the Ivalon patch over the septal defect remain in place or cause later trouble? Will stenosis recur at the point of resection of the infundibular obstruction? Should the outflow tract in the right ventricle be enlarged with an Ivalon graft, and, if so, is there danger of later aneurysm formation? Along with answers to these questions will come more intelligent selection of patients for operation

Those who have been instrumental in developing the shunt operations and infundibulectomy have been accused of being biased in their


Infundibular stenosis
Aortic valves
I V septal defect
Ivalon reconstruction of septal defect
Ivalon reconstruction ventricular wall
Infundibulectomy

favor It has always been true that a father loves and favors hıs own chıld even though the chıld ıs definıtely retarded In self-defense and ın defense of Smıth, Blalock, Taussıg and Brock, I am sure I may say that we are not grıevıng as the ıncomplete operatıons for tetralogy of Fallot are beıng laıd asıde for an operatıon whıch wıll cure and be relatıvely safe As future surgeons pass the markers of these operatıons all we ask ıs that they say a few kınd words

Tricuspid Atresia

Children with tricuspid atresia are always blue. Caval blood can leave the right atrium only by way of an interatrial septal defect into the left atrium, where it mixes with oxygenated blood. The mixed blood enters the left ventricle and is pumped into the aorta and into a smaller than normal pulmonary artery by way of an interventricular septal defect. In those few cases in which the ventricular septum is intact life is maintained by blood shunted through a patent ductus arteriosus. The degree of cyanosis is in direct ratio to the size of the interventricular defect and to the caliber of the pulmonary artery. Patients with tricuspid atresia may be divided into two overlapping groups: those who have little pulmonary blood flow and require emergency surgical relief during early infancy, and those who have sufficient blood flow to the lungs to per[illegible] delay of operation until an opportune time.

favor It has always been true that a father loves and favors his own child even though the child is definitely retarded In self-defense and in defense of Smith, Blalock, Taussig and Brock, I am sure I may say that we are not grieving as the incomplete operations for tetralogy of Fallot are being laid aside for an operation which will cure and be relatively safe As future surgeons pass the markers of these operations all we ask is that they say a few kind words

a pessimistic but energetic resident obtained permission for postmortem examination the day before operation The parents faithfully bring back this boy now twelve years old for a check up examination each year His heart is not much enlarged and he looks and acts like a normal child, although he is somewhat undersize and underweight Cyanosis is visible only slightly in lips and finger tips At each return visit I stare at this child in wonder and as I mentally picture the erratic course of blood flow through his heart I am again impressed by the toughness of cardiac muscle.

Diagnosis

A diagnosis of tricuspid atresia at any age is usually easy to make on the basis of a few specific findings. A child, cyanotic since birth, who has a scratchy systolic murmur at the base of the heart, whose lung fields are clear roentgenographically, whose heart shadow is rounded on the left by hypertrophy of the left ventricle and flattened on the right in the region of the diminutive right ventricle, and whose electrocardiographic tracing shows deviation of the axis to the left—that child has tricuspid atresia.

Treatment

The treatment is surgical. More children require operation early in life for tricuspid atresia than for any other congenital heart deformity. The arch of the aorta in these patients usually descends on the left side. Creation of an artificial ductus by an anastomosis between the left pulmonary artery and the aorta is the operation of choice. The size of the anastomosis varies from 3 mm. in diameter, in infants, to 4 mm. in older children. Occasionally the aorta is so small in tiny infants that the Potts-Smith aortic clamp cannot be used without completely occluding the aorta. In such cases the left pulmonary artery and a lip of the aorta are clamped together longitudinally in a modified curved ductus clamp. Parallel incisions are made in the vessels pinched together in a single clamp, and an aortic-pulmonary anastomosis is performed—not without some technical difficulty. The smallest child we have operated upon with this technique was a 13-day-old infant who weighed 4 pounds, 13 ounces. She survived.

Some surgeons advocate enlargement of the interatrial defect as an essential part of the operation. We have not followed this advice, although the idea is sound. Usually these infants are such poor risks that the extended surgery increases mortality and offsets the possibility of a better end result. Older children do not require enlargement of the septal opening.

Results following operation are rather good, considering the age and condition of the patients. Of 48 infants below one year of age upon whom aortic-pulmonary anastomoses were performed, 38 survived—a mortality of 20 per cent. Twenty of these infants were less than two months old at the time of operation. There was only one death in 25 patients, or a mortality rate of 4 per cent, in children who were one year or older at the time of operation.

If the child survives without heart failure for a month or two following operation, the prognosis is fairly good. A 4-month-old child, deeply cyanotic, in and out of oxygen since birth, had an aortic-pulmonary anastomosis in 1947. No one expected the infant to survive operation, in fact,

Transposition of the Great Vessels

Complete correction of transposed vessels at the base of the heart and closure of the associated intracardiac defects are impossible at present because of the anatomic location of the origin of the coronary arteries Until someone can devise a method of moving the ostia of the coronary arteries along with the aorta, or of rerouting the entire venous circulation cardiac surgeons will have to be satisfied with less than complete correction of this deformity

Diagnosis

The infant without respiratory defects who is cyanotic at birth, and remains so throughout the neonatal period, probably has transposition.

anomalies of the heart and great vessels it is prudent to forewarn parents that unanticipated deformities may prevent accomplishment of the planned procedure

A history of episodes of heart failure is not a contraindication to operation In fact it is not uncommon for children with transposition to have had one or more attacks of decompensation Many children have been operated upon who had a history of congestive failure and at the time of operation had an enlarged liver and some ascites Obviously uncorrectable decompensation is a contraindication to operation. What a child s heart will tolerate is constantly a source of amazement

Operation

During the past ten years a number of operations for relief of cyanosis due to transposition have been tried and discarded In 1956 Baffes presented a new method for partial surgical correction of trans position by shunting all the venous blood from the inferior vena cava to the left side of the heart and the oxygenated blood from the right lung to the right side of the heart. Because these children have decreased platelet counts often as low as 5000 to 10 000 per cubic millimeter it is essential to secure blood less than twenty four hours old for the operation and the immediate postoperative period. Important steps in the operation are briefly as follows

The anesthetized child is laid on the operating table in the left lateral position, and the chest is opened through a posterolateral incision made through the right fifth intercostal space The pulmonary arterial pressure is immediately determined by palpation and, if uncertain by manometer usually palpation alone is enough to determine whether the pressure is high. If the pressure is below 200 mm of water the causative pulmonary stenosis precludes performance of a shift of venous circulation and allows only a subclavian pulmonary anastomosis to relieve cyanosis Pressures above 200 mm of water indicate that the anticipated operation may be performed.

A curved coarctation forceps is applied to the side of the inferior vena cava, and to the excluded lip a segment of homologous aortic graft is sutured

The right pulmonary artery and main stem bronchus are then snugly occluded with umbilical tape to prevent congestion of the lung while the right pulmonary veins are dissected free at their point of entrance into the left atrium. A curved toothed coarctation clamp is applied to the edge of the left atrium at such a level that the pulmonary vein can be cut at a point where it is a single vessel. (Just before this clamp is applied the right pulmonary artery is released for five minutes while the bronchus remains occluded During this time trapped air in the right lung is

The open ductus arteriosus usually shunts enough blood to the lungs to minimize cyanosis for a few weeks or more in infants who have other congenital heart defects, such as tetralogy of Fallot

For the baby who is constantly deeply cyanotic even in an oxygen-flooded Isolette, who has little or no murmur and an enlarged heart, no immediate operation is possible Such infants usually have, besides transposition of the aorta and pulmonary artery, insufficiently large septal defects to allow adequate mixing of oxygenated with unoxygenated blood Observation for a month or more is essential to determine eligibility for later operation Parental pressure upon the attending physician to do something about the cyanosis is apt to force immediate transfer of the baby to a cardiac center The infant is better left in the hospital where he was born and observed for a time to determine the severity of the deformities

Diagnosis in children beyond six months of age is not difficult in typical cases A history of continuous unchanging cyanosis since birth is of prime importance (A few children with a transposition complex show minimal cyanosis) At age two to four years cyanosis is deep, the scleras are blood-shot, and the fingers and toes are black and clubbed Probably because of marrow hyperplasia, these children often have a peculiarly shaped "square" head The unhappy child is undersize and underweight, and has little energy A systolic murmur rather typical of a ventricular septal defect is heard to the left of the sternum at the fourth interspace The heart laboriously slaps against the chest wall with each beat Roentgenograms usually show moderate or great enlargement of the heart, often egg shaped, a narrow mediastinum and clouded lung fields In those few cases in which the diagnosis is in question all doubt is removed by an angiocardiogram which visualizes the aorta arising from the right ventricle

Indications for Operation

It is doubtful whether, in the light of our present knowledge, surgical correction of transposition should be attempted before the infant is at least a few months old Although one six-week-old child did survive operation and has shown satisfactory improvement, it seems preferable to delay operation until there is a reasonable chance for success Parents sometimes force our hands

The child who has transposition of the great vessels, partial transposition or the Taussig-Bing syndrome is a candidate for surgical correction, provided cyanosis is part of the clinical picture If the oxygen saturation of arterial blood is above 75 per cent, the Baffes operation, designed only to relieve cyanosis, is contraindicated

Because transposition is often associated with other congenital

Portal Hypertension

Portal hypertension in children is a tough problem Too few cases are seen in any one institution to allow establishment of fixed rules for management. What is written in this chapter stems from our experience during the past twelve years with twenty four operations performed on eighteen patients. Besides the operative cases we have observed a con siderable number of patients with varying degrees of portal hypertension —some too ill to tolerate operation and others with symptoms too mild to warrant it. Only impressions not conclusions, can be offered.

Etiology

The common causes of portal hypertension in children are (1) cavernous hemangioma of the portal vein, (2) thrombotic obstruction of the vein secondary to ascending infection from neonatal omphalitis Recanalized thrombosed veins may be indistinguishable from heman

absorbed, and the lung collapses and allows more room to work) The proximal end of the pulmonary vein is left open to discharge blood that may have accumulated in the lung

The other end of the graft previously anastomosed to the side of the inferior vena cava is now sutured to the stump of the pulmonary vein on the left atrium The other end of the pulmonary vein is sutured to the right atrium Ligatures and clamps are released, and the lung is re-expanded After the child has stabilized, the inferior vena cava is tied with a strong silk ligature between the point of attachment of the graft and the right atrium This complete occlusion makes obligatory the entire blood flow from the inferior vena cava through the graft into the left atrium

RESULTS. In spite of the fact that only 60 per cent of the venous circulation is rerouted to the *left* atrium and a nearly similar amount of oxygenated blood is directed to the *right* atrium, these children do remarkably well after operation They are able to run, swim, ride bicycles, and enjoy other moderate activities Oxygen saturation of arterial blood rises from the usual preoperative level of 55 to 65 per cent to 85 to 90 per cent Obviously, some degree of cyanosis persists, but often when the patient is at rest it is barely visible

One hundred and twenty-one children, ranging in age from six weeks to twelve years, have been operated upon at the Children's Memorial Hospital for transposition of the great vessels during the past three and one-half years Operative mortality remained at approximately 30 per cent in the first 85 cases Better surgical technique, use of fresh instead of preserved blood, and more discerning postoperative care have slowly reduced the hazard of this rather extensive operation During 1958 and two months of 1959, 36 patients were operated upon with only 4 deaths, or a mortality of 11 per cent

The operation is far from perfect, but is the best one available to date Experimental work is being done on refashioning the right atrium so as to reroute the entire venous circulation

Surgical Treatment

It is always difficult to know when to operate and what type of operation to perform We do not consider a single hemorrhage sufficient indication for operation although it well may be After a subsequent hemorrhage parents are more amenable to the idea of surgery and the surgeon feels more justified in advising the rather formidable operative procedure This attitude towards indications for operation in case of bleeding is the same whether the child has intrahepatic or extrahepatic portal obstruction

It is difficult to know what to do for the jaundiced child who has bled from esophageal varices If the thymol turbidity and cephalin flocculation tests indicate severe impairment of liver function and if the albumin globulin ratio is reversed operation has little to offer Our medical confreres are apt to urge operation in anticipation of eliminating hemorrhage If a period of attentive medical management results in improvement of the child s general condition and some reversal of the ominous liver pathology operation is performed, but with the definite understanding that the prognosis still is poor Operation is contraindicated for those children whose liver pathology is irreversible.

Three basic operations are available for the relief of portal hypertension portacaval anastomosis splenorenal anastomosis and splenectomy alone. In general, it appears that a portacaval anastomosis is the first choice, especially for children below three to five years of age.

Splenorenal anastomosis is acceptable for the older child with a large spleen The greatest advantage of a portacaval shunt lies in the fact that a large anastomotic channel can be formed Undoubtedly a portacaval shunt is less apt to become occluded by thrombosis than is a splenorenal shunt. It is questionable whether a splenorenal anastomosis will remain open in a child below five years of age because of the small caliber of the splenic vein.

Seven years ago a splenorenal anastomosis was performed on a two-year-old child who had had two bouts of hematemesis had a large spleen and was slightly jaundiced. From microscopic studies of a liver biopsy a diagnosis of hepatitis was made This child is well and hasn t bled since, probably not because of the shunt, but because the hepatitis resolved and the liver regenerated.

Splenectomy is beneficial when the splenic vein is occluded by thrombosis Occasionally the sequelae of portal hypertension are relieved by splenectomy alone even though the spleen is not large and all its vessels are open

PORTACAVAL ANASTOMOSIS. The child is placed on the operating table on his left side in a semilateral position. Under intratracheal anesthesia a low transthoracic, transrectus muscle incision is made To

giomas (3) Cirrhosis of the liver secondary to hepatitis or liver poisoning. The causes of portal hypertension in our experience have been equally divided between intrahepatic and extrahepatic pathology

Symptoms

Massive gastrointestinal bleeding from esophageal or gastric varices is the primary complication of portal hypertension Without premonitory signs or symptoms the child with *extrahepatic* obstruction suddenly vomits large amounts of bright red blood or passes tarry stools Bleeding may occur at any age, six of our patients were between nine months and three years when the first episode of bleeding occurred

The child who has been well until the time of hemorrhage usually has a normal liver The spleen may or may not be enlarged. Ascites is not common, but does occur Jaundice is rare

Some years ago a mother reported that her child, with known portal hypertension, was most apt to have a hemorrhage during an upper respiratory tract infection This association has since been noted frequently enough to suggest more than coincidence It seems reasonable to suppose that swallowed virulent bacteria may cause local inflammation in the esophagus and rupture of the thin-walled varices

Jaundice of some degree, usually mild, is often the first sign of *intrahepatic* obstruction Hepatomegaly, splenomegaly, secondary anemia, neutropenia, a tendency to gastrointestinal hemorrhage and, in the latter stages of cirrhosis, ascites and deepening jaundice characterize intrahepatic obstruction of the portal vein Although the term "Banti's disease" has been largely discarded, his description of the syndrome is still applicable

Treatment of Hemorrhage

Sudden vomiting of large amounts of blood and/or passage of copious tarry stools demand hospitalization and adequate blood transfusions As the blood pressure falls bleeding usually stops spontaneously In those few instances in which bleeding continues, use of a children's size Blakemore balloon as a tamponade in the esophagus is lifesaving The triple tube with balloon attached is passed through a nostril into the stomach The smaller balloon is inflated and drawn against the cardia of the stomach, where it is held by continuous mild traction The larger intraesophageal balloon is attached to a blood pressure apparatus, and inflated to pressure of 30 mm of mercury and maintained at that level An aspirator is attached to the third tube to keep the stomach empty

Pressure necrosis may occur if the balloons are left in place more than twenty-four to thirty-six hours

Surgical Treatment

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PORTACAVAL ANASTOMOSIS The child is placed on the operating table on his left side in a semilateral position Under intratracheal anes thesia a low transthoracic, transrectus muscle incision is made To

provide adequate exposure, the liver is displaced upward through a generous incision in the diaphragm Portal pressures are taken and usually found to be between 300 and 400 mm of water The portal vein is isolated, if it is thrombotic, nothing further can be done In one instance we isolated a mesenteric vein and sutured it to the inferior vena cava, but without subsequent benefit to the patient In another instance with much difficulty a bulge of a cavernous hemangioma of the portal vein was isolated in a fourteen-year-old boy and successfully sutured to the cava He is now twenty-four years old and has remained well and free from hemorrhage

A normal-sized portal vein is anastomosed to the side of the inferior vena cava If the anastomosis can be made below the origin of the renal veins, a large aortic clamp is placed around the cava and a lip of the vessel occluded The portal vein is drawn to the aortic clamp and completely occluded Long incisions are made in both vessels to provide the largest possible anastomotic channel When the portal vein can be brought to the cava only at a level with the renal veins, a lip of the cava and the portal vein are simultaneously occluded with a curved toothed coarctation forceps A strip of vena cava is excised to provide a large channel and lessen the chances of thrombosis and obstruction

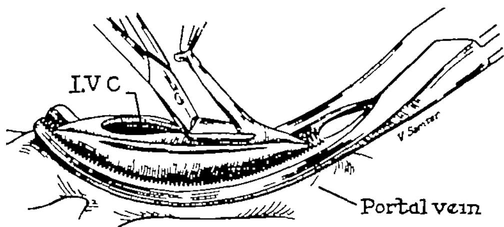
SPLENORENAL ANASTOMOSIS. The child is placed in the opposite position to that for a portacaval anastomosis A similar large incision is made, and portal pressures are measured The splenic artery is doubly ligated and cut The splenic vein is carefully dissected free, occluded with a toothed ductus forceps and divided at the point where it flutes out as it breaks up into branches The spleen is removed

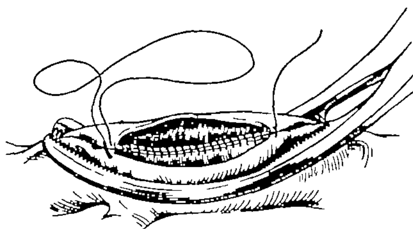
The renal vein is thoroughly isolated, and a longitudinal lip is occluded with a curved toothed coarctation forceps in such a manner that the renal flow is not completely obstructed The renal vein is opened, and the end of the splenic vein is sutured to it with a running suture of 6-0 Deknatel silk on a nontraumatic needle, the finer the suture material, the less the chances of thrombosis

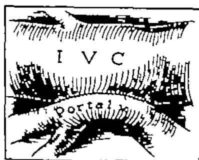
A biopsy of the liver is always taken

RESULTS Twenty-one portacaval or splenorenal shunts were performed on eighteen patients Splenectomy alone, suture of the esophageal varices, and resection of the esophagus and fundus of the stomach were each performed once

Five patients died at various periods after operation, four of cirrhosis of the liver and one four years after operation of overwhelming infection and bilateral hemorrhage in the adrenals Two of these patients had advanced cirrhosis of the liver at the time of operation Four patients remain cured from five to ten years Because of recurrent bleeding a second operation has been necessary in four patients, they are doing well, but a sufficient number of years have not passed to claim cures







Completed
anastamosis

Another child who had a splenorenal anastomosis in 1952 at age five years has recently bled Injection of the varices in the esophagus with sclerosing solution has been unsuccessful, he will need a portacaval anastomosis One child could not be traced The remaining three patients have been operated upon within the last year and cannot be evaluated, although they have had no recurrent hemorrhage to date

To emphasize the gravity of this problem of portal hypertension and to illustrate our lack of knowledge about this condition, case reports of two children are briefly outlined

An eight-month-old boy was admitted to the hospital because of a second episode of severe hematemesis There was a history of postnatal omphalitis His liver profile was normal At age nine months a splenectomy was done with the faint hope that this might be of temporary benefit—it wasn't Repeated hemorrhages followed, each requiring blood transfusions At age one year a portacaval anastomosis was performed, also without benefit A number of uncontrollable hemorrhages required insertion of the Blakemore balloon At age two years the lower end of the esophagus was opened, and the varices were sutured Severe hemorrhages persisted At age two and one-half years the lower end of the esophagus and all the acid-bearing portion of the stomach to the antrum were resected An esophagogastrostomy was performed The boy is now four years old During the eighteen months since the last operation there has been no hematemesis, but an occasional dark stool The boy is well, but not robust, his appetite is finical, and he looks pale and has less than normal energy Every time I see him I renew my vow never again to do a radical stomach resection on a child In retrospect, it appears that it would have been wiser to replace the resected lower end of the esophagus and a portion of the fundus of the stomach with a transplanted segment of bowel as suggested by Merendino

An eleven-month-old boy, weighing fifteen pounds, was admitted to the hospital because of hematemesis and severe ascites His liver profile too was normal A portacaval anastomosis was attempted, but found to be impossible because of complete occlusion of the portal vein Hemorrhage recurred, and weekly abdominal paracenteses were necessary At age fourteen months, in desperation, a splenectomy was advised The spleen, consisting of four individual masses each about golf-ball size, was removed After this operation the child simply got well Neither ascites nor hemorrhage has recurred He is now seven years old and as normal as any child his age, except for long transverse scars on each side of his abdomen Why did he suddenly get well?

From these cases and a hodgepodge of experience no conclusions can be drawn At the moment we are inclined to believe that for control of bleeding from esophageal varices due to portal hypertension in children, portacaval anastomosis offers the best chances of success

Congenital Atresia of the Bile Ducts (Jaundice)

Congenital atresia of the bile ducts is the darkest chapter in pediatric surgery. The etiology is unknown; both prenatal infection and congenital maldevelopment have been considered. Infection is the more attractive theory because it offers hope of finding the offending agent. In the light of our present knowledge, unless bile can be shunted to the gastrointestinal tract, early death is inevitable.

Jaundice

Jaundice during infancy is common. Physiologic jaundice appearing a few days after delivery and persisting for a week or more is incon-

sequential except occasionally in a premature infant Bile is always present in the stool The jaundice disappears in a week or two The liver remains undamaged

Jaundice following erythroblastosis fetalis is caused by excessive destruction of red blood cells, appears promptly after birth and is recognized as such by well established laboratory tests The jaundice is usually deep, and persists for a week or ten days in treated patients and for a few weeks to several months in the untreated In some patients with erythroblastosis the bile ducts become obstructed by "inspissated bile," a descriptive term coined by Ladd Whether the concentration of bile is due to liver cell damage or to excessive accumulation of bilirubin pigment in the ductal system, or both, is unknown Eventually the jaundice spontaneously clears and the liver remains free from degenerative changes

Serum and viral hepatitis in infants has been recognized as a common cause of jaundice for the past ten years The increasing number of reports of cases indicate that hepatitis during the neonatal period is not uncommon Jaundice may appear during the first or second week after delivery and is apt to be thought due to atresia of the bile ducts It seems reasonable to assume that many cases of unexplained jaundice and many cases of cirrhosis of the liver appearing during early or late childhood are due to serum or viral hepatitis

Whether jaundice may be due to inspissated bile as an entity—not that which follows erythroblastosis—is a matter of dispute at present After the common causes of jaundice have been eliminated there remain a number of cases which fall into no specific diagnostic category They are said to be due to inspissated bile or a mucus plug in the common duct Whether inspissated bile in these indefinable cases is the cause of jaundice or is the result of mild hepatitis, cholangitis, congenital narrowing of the extrahepatic ducts, immaturity of the liver or some hitherto unrecognized disease of the liver is uncertain We are inclined to support the claim that inspissated bile and mucus plugs are secondary to some type of basic pathology

Persistent jaundice during infancy is most commonly due to atresia of the external biliary passages The entire ductal system is usually replaced by fibrous cords The few cases in which portions of the common or hepatic ducts are open and available for anastomosis to the duodenum make surgical exploration of all these infants worth while

Diagnosis

Analysis of the causes of jaundice in infants is largely a matter of a careful history, inadequate as it may be, close observation of the child and repeated stool examinations Although the large majority of cases

of jaundice are due to conditions mentioned above, such rare causes as septicemia syphilis toxoplasmosis, cytomegalic inclusion disease, and others, must not be forgotten.

The history of jaundice as given by the mother requires skeptical analysis During the first months of life—except in cases of profound jaundice following erythroblastosis—neither the mother nor the physician is sure how long the child has been jaundiced, nor whether the jaundice has been continuous or intermittent. The obstetrician probably observed a tinge of jaundice considered it physiological and said nothing about it to the mother The mother reports that a week or two after leaving the hospital she noticed that her baby was jaundiced or that attention was called to it by her physician Questions about intermittency of jaundice during the neonatal period (twenty-eight days) usually get affirmative answers because the mother does not know that yellow discoloration of the skin and sclera is poorly discernible by artificial light and that the color of urine is somewhat dependent upon fluid intake

When a four to six week-old jaundiced infant is admitted to the hospital for study innumerable laboratory tests for liver function should be secondary to careful, persistent and intelligent examination of the stools for bile. It seems much easier to order a host of laboratory studies than to examine the stools for bile For some mysterious reason every new house officer must be reminded that examination of a stool passed in a diaper is worthless The baby passes bile-stained urine over the stool and thereby makes the examination worse than useless—actually misleading *The stool to be examined for bile must be taken out of the rectum* Without delay it must be sent up to the laboratory and be examined before it dries up into a little pebble that rattles around in the jar

The color of the stool is not a reliable guide to the presence or absence of bile The baby's diet, medication and the length of time the stool has lain in a urine-soaked diaper all influence the color of the stool and make gross observations misleading. It is well to urge nurses routinely to describe the color of all stools but their notes may be used only as an indicator and not as scientific evidence of the clinical course of jaundice

There are a number of laboratory tests for bile in the stool which if carefully done, are fairly accurate Report of a strongly positive test for bile in the stool is anticipated when the specimen is normally yellow brown. The questionable specimens usually draw a report which reads "A faint trace of bile"—"faint" is largely a matter of opinion. What is sorely needed is a simple test that anyone can do with assurance of accuracy A miniature Nobel prize awaits the investigator who discovers a quantitative and qualitative test for bile in the stool as reliable as the Haines or Fehling solution test for sugar in the urine Procurement of a stool from the rectum twice a day and immediate examination for bile

by a trustworthy test will do much towards accurate diagnosis of jaundice in infants

The two- or three-month-old jaundiced infant who has shown no bile in the stool, who is constantly jaundiced and has been since or shortly after birth, who eats well and manifests no particular signs of illness probably has congenital atresia of the bile ducts For years physicians have been asked whether they could recall the color of the stool of an infant who later was known to have congenital atresia of the bile ducts Thus far no answer has been obtained Medical literature states that the stools of jaundiced infants are acholic, but no single reference can be found which described the stools immediately after delivery Presumably, the first stool should be colorless, but is it? Armchair reasoning suggest that a diagnosis of atresia of the bile ducts could be made at birth if the characteristic of the stools were known We have dozens of tests for liver function, but no recorded observations about such a simple thing as the color of the stool in an infant with congenital atresia of the bile ducts

It is important to differentiate hepatitis from atresia of the bile ducts because operation in the former is valueless and may be fatal Hepatitis may occur at or shortly after birth, but is more apt to appear after a month or two The infant with hepatitis may have fever for a few days at the onset of illness A reliable history of freedom from jaundice for a period of time after birth is of great diagnostic value If the clinical picture resembles that of atresia of the ducts, but some bile unquestionably is found in the stool—even once—the diagnosis is probably hepatitis The history of the mother's pregnancy, of transfusions, and of other cases of hepatitis in the family are important guides to diagnosis It is known that giant cell hepatitis may affect a number of children in one family

Needle biopsy of the liver has been done a number of times when a differential diagnosis was impossible The segment of liver obtained by use of a Vim-Silverman needle is small, but usually sufficient to make a diagnosis A network of fine biliary ducts embedded in fibrous tissue, deposits of bile pigments in the ducts and liver cells, and proliferation of ductal epithelium signify congenital atresia of the bile ducts Evidence of infection and necrosis, and many typical multinucleated giant cells speak for hepatitis

Cholecystograms are useless in cases of atresia of the bile ducts and hepatitis

During recent years transaminase studies have been used to determine the degree of liver destruction Serum glutamic oxalacetic transaminase (SGOT) levels are of value in determining not only the extent of liver injury, but also the degree of destruction In cases of atresia of the bile ducts the transaminase levels remain high A persistent fall in

transaminase levels indicates that the process of liver destruction is being slowed and suggests an improved prognosis This test of liver function is rather popular at present, probably because agile use of the big words suggests erudition.

Before operation plasma protein levels are measured and deficiencies corrected. Other standard liver profile tests such as thymol turbidity and flocculation tests, are of value primarily for prognosis

Treatment

When to operate upon a child who has atresia of the bile ducts is still disputed. Arbitrarily we have chosen two months of age as the most suitable time—if we have a choice. Irreparable liver damage does not occur in two months or even three However after six months of complete jaundice the liver is hard and fibrotic and cannot be expected to return to normal if operative correction should be possible.

Before operation a few doses of vitamin K—not more than 2 mg per dose—are given parenterally Too much vitamin K is toxic. Blood is given as indicated.

The infant is placed on an x ray plate on the operating table To avoid lost time, confusion and breaks in surgical technique, it is advisable routinely to prepare in advance for x ray examination during operation on any jaundiced infant or child

Under open-mask ether anesthesia a right upper quadrant incision—longitudinal or transverse—is made At times when the diagnosis rests between atresia of the ducts and hepatitis, but is inclined to the latter a small incision is made. The gallbladder is exposed. If it contains nothing but white mucus—there is no such thing as white bile—a diagnosis of atresia is established. However if the gallbladder contains bile the course of the operation is immediately changed. With a fine needle approximately 10 cc. of 30 per cent Diodrast is slowly injected into the gallbladder Demonstration of contrast medium in the duodenum is incontrovertible evidence that the bile passages are patent and that no further surgery is indicated

When no bile is found in the gallbladder—the majority of cases—a thorough, painstaking search is made for the common and hepatic ducts Sharp dissection with a fine scissors is most satisfactory The portal vein and hepatic artery are identified and isolated. Close attention to hemostasis and occasional lavage of the operative field with salt solution are helpful for the identification of structures The hilus of the liver is meticulously explored. Any bulging structure suggesting a possible dilated duct is aspirated with a fine needle, it usually turns out to be a dilated vein No structure which might be a remnant of a hepatic duct is left unexplored by needle or scissors After the dissection is complete

by a trustworthy test will do much towards accurate diagnosis of jaundice in infants

The two- or three-month-old jaundiced infant who has shown no bile in the stool, who is constantly jaundiced and has been since or shortly after birth, who eats well and manifests no particular signs of illness probably has congenital atresia of the bile ducts For years physicians have been asked whether they could recall the color of the stool of an infant who later was known to have congenital atresia of the bile ducts Thus far no answer has been obtained Medical literature states that the stools of jaundiced infants are acholic, but no single reference can be found which described the stools immediately after delivery Presumably, the first stool should be colorless, but is it? Armchair reasoning suggest that a diagnosis of atresia of the bile ducts could be made at birth if the characteristic of the stools were known We have dozens of tests for liver function, but no recorded observations about such a simple thing as the color of the stool in an infant with congenital atresia of the bile ducts

It is important to differentiate hepatitis from atresia of the bile ducts because operation in the former is valueless and may be fatal Hepatitis may occur at or shortly after birth, but is more apt to appear after a month or two The infant with hepatitis may have fever for a few days at the onset of illness A reliable history of freedom from jaundice for a period of time after birth is of great diagnostic value If the clinical picture resembles that of atresia of the ducts, but some bile unquestionably is found in the stool—even once—the diagnosis is probably hepatitis The history of the mother's pregnancy, of transfusions, and of other cases of hepatitis in the family are important guides to diagnosis It is known that giant cell hepatitis may affect a number of children in one family

Needle biopsy of the liver has been done a number of times when a differential diagnosis was impossible The segment of liver obtained by use of a Vim-Silverman needle is small, but usually sufficient to make a diagnosis A network of fine biliary ducts embedded in fibrous tissue, deposits of bile pigments in the ducts and liver cells, and proliferation of ductal epithelium signify congenital atresia of the bile ducts Evidence of infection and necrosis, and many typical multinucleated giant cells speak for hepatitis

Cholecystograms are useless in cases of atresia of the bile ducts and hepatitis

During recent years transaminase studies have been used to determine the degree of liver destruction Serum glutamic oxalacetic transaminase (SGOT) levels are of value in determining not only the extent of liver injury, but also the degree of destruction In cases of atresia of the bile ducts the transaminase levels remain high A persistent fall in

were normally formed, but hard as cords. The gallbladder contained no bile. The common duct, about 3 mm in diameter was opened by a longitudinal incision A lumen could not be identified, but a layer of shiny cells near the center of the duct suggested that one might be present. A fine probe was introduced and with some maneuvering could be pushed well up into the right hepatic duct in the liver No bile appeared when the probe was withdrawn The abdomen was drained and closed On the third day after operation copious amounts of bile began to flow from the wound. After six weeks of daily drainage of 300 to 500 cc. of golden yellow bile, the abdomen was reopened in anticipation of finding a duct and suturing it to the duodenum. Because identification of a duct in the mass of fibrous tissue was impossible a Roux Y anastomosis of the jejunum to the liver was performed. After operation bile promptly appeared in the stool. The child is thriving, but has occasional attacks of jaundice which spontaneously clear The child s future is uncertain but temporarily she has been helped.

In 1946 and 1947 three unusual cases of clearance of jaundice coincidental with operation almost led us to premature publication of a paper Three infants who presumably had been continuously jaundiced were operated on because of a diagnosis of atresia of the bile ducts At operation in each instance a normal biliary tract was found, but at the junction of the common duct with the duodenum lay a large lymph node which kinked and apparently obstructed the common duct. After removal of this gland, bile appeared in the stool. In retrospect, most likely these were cases of hepatitis not recognized as such at the time. Whether removal of the lymph node was instrumental in relieving jaundice or whether it was a matter of pure coincidence is unknown. Oddly enough we have not had a similar experience since

The majority of children with congenital atresia of the bile ducts die between their first and second years a few live four or five years. Eventually the liver becomes hard, the abdomen fills with fluid, and the child ceases to eat and without much suffering fades away One would expect itching to be a problem in these deeply jaundiced patients but it rarely is The occasional child who is troubled may be relieved by aspirin given at regular intervals

It is more than coincidence that these children although jaundiced and presumably always somewhat uncomfortable, are generally happy and smiling. This is pure assumption maybe these infants are so well dispositioned because of the extra care and love they receive Parents know that their child will not survive and consequently shower him with affection The response is a well adjusted child It s only an observation, but the principle might be worth application to the irritable and ill-dispositioned child

any depression in the hilus of the liver is explored with a probe Since the prognosis is hopeless in all cases in which no duct is found, the surgeon should use no half-measures in the exploration Nothing can be lost in opening any area which might contain a miniature bile duct

In the rare instance in which the common duct is atretic, but the gallbladder contains bile, an anastomosis between it and the duodenum is performed with interrupted 5- or 6-0 silk sutures on a swaged-on needle An open stump of common or hepatic duct is anastomosed to the duodenum The posterior edge of the duct is fixed to the side of the duodenum with a few interrupted sutures Then one end of a 1-inch long, properly fitting plastic tube is inserted into the common duct and the other end into the duodenum The anastomosis is completed over the plastic tube Fine silk on a swaged-on needle is essential to avoid fraying the end of the bile duct. The small tube will be discharged into the bowel in a few days

A biopsy of the liver is routinely taken

It is discouraging to be able to do nothing for infants in whom no remnant of bile ducts can be found About ten years ago we had an idea that bile ducts might be induced to form by making multiple cuts in the liver substance Like many good ideas, it didn't work The valvulotome used for opening a stenotic pulmonary valve was inserted in the closed position into the hilus of the liver The blades were then expanded, and multiple cuts were made in various directions well into the substance of the liver and presumably across small bile ducts Bleeding was minimal A catheter, perforated with many side holes, was then inserted into the opening in the under surface of the liver The distal end of the catheter was brought to the outside through the upper end of the incision It seemed reasonable that opened ducts might discharge bile and thus form a fistula to the outside After operation the catheter was kept open by flushings with salt solution and weak peroxide This procedure was tried in six cases, but in no instance did bile appear in the test tube hopefully attached to the catheter

From time to time reports appear in medical literature of relief of jaundice following anastomosis of the small bowel to a scarified area of liver or to the resected end of the left lobe of the liver in infants with atresia of the bile ducts It seems altogether reasonable to assume that coincidental with such an operation an overlooked common duct spontaneously opened and took over its normal function

RESULTS The surgical treatment of congenital atresia of the bile ducts is most discouraging During the past twelve years approximately sixty patients have been operated upon, and only three could be cured A few were helped temporarily This case is illustrative At three months of age a child who had been continuously jaundiced was subjected to exploratory operation The gallbladder, common duct and hepatic ducts

Umbilical Hernia

An umbilical hernia is often just an incidental finding to the pediatrician, but a problem of deep concern to the mother

An umbilical hernia is a weak spot in the mid-abdominal fascia where the blood vessels entered and left the abdomen during fetal life For some unknown reason it is more common in colored than white children Umbilical hernias come in all sizes from the tiny openings which will barely admit the tip of one's little finger to those which will easily admit two or three fingers The small hernia often contains only a nubbin of properitoneal fat, the large hernia, collapsed when the child is at rest, is widely distended by loops of bowel and protuberant when the child cries All are symptomless and practically never become incarcerated in children. Our staff has met with only two incarcerations over a period of many years and unusual circumstances attended both One child had a peach pit caught in a loop of bowel protruding through the umbilicus the other had a hard piece of feces caught in a similar position.

delay of operation is not dangerous or indicative of neglect and that the hernia may close spontaneously It often is harder to talk parents out of than into an operation on their child.

The operation itself is a simple one. Under ether anesthesia a small curved incision is made in the loose skin of the umbilicus The peritoneal sac is dissected from the fascia and closed by a purse string or running suture of surgical gut. The opening in the fascia is closed transversely with an appropriate number of silk stitches To form a normal appearing inverted umbilicus, a silk stitch is placed in the under surface of the most protuberant skin of the umbilicus and is brought down and stitched to the anterior surface of the fascia. The skin is closed with a couple of fine subcuticular sutures A little piece of gauze is wadded up placed over the umbilicus, and fixed with adhesive tape The child leaves the hospital the same day or the following morning.

Probably one of the most unkind things a surgeon can do is to remove the umbilicus When the child grows up he or she will be sensitive about the wide expanse of abdomen with no landmark.

The question of what to do with an umbilical hernia has been a matter of sharp dispute for many years. Some claim that none should be treated surgically, others maintain that operation is the only effective cure. As in most disputes, it appears that both sides are half-right. The tiny hernia which will admit only the tip of one's finger will close spontaneously. In an infant less than one year old the hernia which will admit one's index finger will probably close. It seems doubtful whether the large umbilical hernia which constantly contains loops of bowel will close satisfactorily.

If we agree that the large hernia should be operated upon and that the little ones should not, we are still left with a sizable number of questionable cases which require some attention, or at least simulated attention. The mother sees the hernia protrude every time the child cries and is sure that the bluish skin will some day rupture. She demands that something be done. The accepted treatment for as many years as we have had adhesive tape is strapping. Before consulting their doctor parents have often been strapping the abdomen according to directions of neighbors or relatives. Buttons, nickels, wads of gauze, pebbles, all sorts of objects are plastered over the hernia with adhesive tape. Learned textbooks give detailed advice about how adhesive tape placed on the skin painted with tincture of benzoin should be drawn up so tightly that the hernia is kept reduced. The advice is fine, but the execution leaves something to be desired. In a few days the adhesive slips a bit and the hernia again protrudes. Or the adhesive is put on so tightly that the skin gets sore and in a few days has to be removed.

A number of years ago it seemed worth while to try to determine whether adhesive strapping actually did any good. Every other child in the out-patient department with an umbilical hernia was strapped. The mothers whose children received no treatment felt slighted, but were mollified by a bit of pure quackery. The babies of the control series had their umbilical areas painted with picric acid. The brilliant yellow color of the solution remained on the skin of the control group about as long as the adhesive stuck in the other group, and all were happy. The war came on before this profound research could be completed, but during the period of observation no difference could be observed between the two groups. Some do not think that adhesive strapping does any good, and I'm inclined to hold to that idea, but of this I am convinced. If it appears that the hernia may close spontaneously, it is wise to have the process attended by some adhesive over the skin, or the mother will find a wiser doctor.

So long as indications for surgical repair are indefinite, each case will have to be decided upon its own merits. One will appropriately and justifiedly feel a bit more virtuous if operation is put off a number of months in the borderline cases and it is explained to the mother that

Omphalocele

For a short time during early embryonic life the rapidly growing, primitive bowel pushes itself out of the abdominal cavity If it fails to return, the infant is born with an omphalocele The bowels protruding through the umbilical region are encased in a thin white fragile membranous sac made up of peritoneum fused with a layer of amniotic membrane

The extent of herniation determines the size of the defect in the abdominal wall. Small herniations of a few loops of bowel through a defect 2 to 4 cm. in diameter are as easily repaired as an umbilical hernia and will not be discussed. Herniations of most of the intestine and a considerable portion of the liver through abdominal defects 6 to 10 cm. in diameter present vexing problems

Every omphalocele is obvious at birth. Because the herniation is actually into the umbilical cord itself the diagnosis cannot be missed as the cord is being tied.

hesions. Before Gross made the suggestion of retaining this membrane we excised it and as a result at the second operation struggled with fibrous adhesions between the under surface of the skin and the bowel.

In those cases in which the undeveloped abdominal cavity will not completely contain the viscera and it is possible to cover them only with skin, an attempt still must be made to bring the rectus muscles together at either the upper or lower angle of the wound The importance of apposing even in an inadequate fashion some portions of the rectus muscles cannot be overemphasized The short laterally placed rectus muscles will not be stretched if the viscera push themselves forward through the opening in the abdominal wall against the easily yielding elastic skin But if the muscles have been drawn over part of the viscera, they will stretch and at a later operation can be completely drawn together

At what age the second attempt at closure of the abdominal wall should be made is a matter of opinion We believe the second attempt should be made within a few weeks or less depending upon the condition of the child The longer one waits the more the abdominal viscera protrude and the more the rectus muscles are pushed laterally Eventually it becomes impossible to bring them over the top of the mushrooming viscera. We have seen four older children whose bowels literally were hanging in a sac Repeated attempts two in one child and three in another to bring even a portion of the muscles over the viscera met with failure

When closing or attempting to close the abdomen for an infant with a large omphalocele, some philosophical judgment will have to be exercised. Pulling the muscles together under much tension over the protruding liver and bowels may lead to respiratory distress because of pressure on the diaphragm, and to circulatory impairment of the legs due to excessive pressure on the systemic circulation in the abdomen What should one do? Two courses are open one may take out the muscle sutures simply close the skin and save the baby's life, recognizing at the time that later closure will almost certainly be impossible and that the child therefore will grow up under a rather serious handicap Or one may leave in the muscle stitches and hope that the baby survives to grow up with no handicap How many chances may a surgeon take with life for the sake of restoring a child to normal and assuring an opportunity for happiness?

Management

If the sac is intact, it should immediately be lavaged with Zephiran solution and covered with gauze soaked in saline solution If the sac has ruptured during delivery, the exposed viscera should be irrigated with copious amounts of saline solution to remove bits of meconium and any contamination that may have occurred during delivery The loops of bowel are then encased in an envelope of moist gauze and covered with a towel encircling the abdomen In those few instances in which the sac ruptured some time before delivery the bowel surfaces will be red and matted together by fibrous adhesions as a result of intrauterine chemical peritonitis

Whatever the local condition, immediate operation is indicated Every hour of delay increases the danger of infection and, because of rapid distention of the intestine with air, makes the operation more difficult However, before rushing to surgery it is well to examine the infant rather carefully and have a roentgenogram of the chest because no other group of infants has a higher percentage of associated anomalies Even though repair of the omphalocele is urgent, it is well, if for no other reason than for the surgeon's protection and peace of mind, to know of other deformities and inform the relatives of the added seriousness of the child s condition One always feels a bit chagrined to have the postmortem examination bring to light a second or third congenital anomaly which might have been discovered by routine examination

Surgical problems which will be encountered are in direct ratio to the extent of herniation Actually, the cases can be divided into two groups those in which the liver protrudes through the abdominal wall and those in which it does not The liver is easily recognized by its dark blue color shining through the translucent sac Rarely will any difficulty arise in closing the defect if the liver does not protrude

Large omphaloceles containing much of the bowel and a portion of the liver require some analytical thought before beginning the operation Is it going to be possible to replace the viscera and close the abdominal wall? If the surgeon is convinced that it will be possible, the sac with a small strip of skin around it is excised, the viscera are replaced, and the abdominal wall is closed in layers with fine interrupted silk or cotton sutures If there is the slightest uncertainty, however, about the possibility of accomplishing these objectives, a different surgical attack is in order An incision is made through the skin around the sac, but the sac itself is left intact Should it be impossible to return the viscera to the abdominal cavity and close the abdominal wall, no choice remains but to free the skin widely on each side and use it for closure The importance of preserving the sac is apparent it serves as a smooth protecting membrane between the bowel and the skin and prevents subsequent ad-

Pyloric Stenosis

The operation for pyloric stenosis is the most satisfactory procedure in the entire field of pediatric surgery The sick baby vomits all its feedings the mother is distraught, a simple operation is performed, the baby thrives and the mother is happy

The infant with pyloric stenosis (usually a boy often the first born) after a normal postnatal period of two to three weeks suddenly begins to vomit all its feedings Although the average age of infants admitted to the hospital with pyloric stenosis is twenty-one days, it is not uncommon to find the condition at age two or three months We have even seen a typical case in a child six months of age.

The vomitus consists of curdled sour formula, it is never green If the vomitus is green the obstruction must be distal to the pylorus and the baby cannot have pyloric stenosis These dictatorial statements were repeated to the house staff for years until a baby with typical pyloric stenosis upset them by persisting in vomiting green material before operation Five years have passed since this exception to the rule was

When a tumor cannot be palpated and the roentgenologist's report is equivocal it is wise to defer operation not only because the child may not have pyloric stenosis but also more important because he may have some other disease causing persistent vomiting A three week-old infant was admitted to the hospital with a classic history of pyloric stenosis We saw large peristaltic waves and thought we felt a tumor A first-class pediatrician concurred Because all the findings were typical I operated upon this child and found a perfectly normal pylorus (We should have heeded the roentgenologist's advice.) A few days later an intravenous pyelogram was made and to our chagrin bilateral hydronephrosis and hydroureters were demonstrated Moral one shouldn't *think* one feels a tumor If one doesn't without doubt feel the tumor further diagnostic studies are indicated

Treatment

Pyloromyotomy is the only treatment for pyloric stenosis The excellent results following operation allow no other conclusion If the infant with pyloric stenosis is admitted in good condition there is little reason for delaying operation However pyloromyotomy is never an emergency procedure Fluid and electrolyte imbalance must first be corrected in the dehydrated child A plastic tube is inserted into the saphenous vein at the ankle, or a needle is introduced into a scalp vein for accurate administration of fluid and blood It is amazing to see how a dehydrated, listless wrinkled infant will respond to such treatment over a period of twenty four hours We believe it is wise to have blood chloride and plasma carbon dioxide determinations before operation is scheduled These needn't be normal to make operation safe but should be reasonably so Clinical judgment is still of great value in choosing the proper time for operation.

After the baby has been placed on the operating table, before anesthesia is begun, a no 16 French catheter is introduced through the mouth into the baby's stomach. With an Asepto syringe accumulated fluid and unsuspected retained formula are removed by aspiration The importance of emptying the stomach cannot be overemphasized. It was made obligatory after this near fatal experience During operation, just after the stomach had been delivered from the abdomen the anesthetist said, "The baby has stopped breathing" It suddenly occurred to me that while pulling up the stomach I might have forced some contained fluid back into the throat and obstructed respiration A lap pad was placed over the wound the sterile drapes were tossed off the infant was grabbed by its legs and held upside down. A little fluid ran out of the baby's mouth, and normal breathing was restored An anxious moment! The operation was successfully completed, and the wound healed without

encountered and we have returned to our dogmatic teaching, occasionally, for the record, we add—with very few exceptions

Diagnosis

A diagnosis is easily made in the textbook case and even in the atypical case if the possibility of pyloric stenosis as the cause of unexplained, continuous vomiting is borne in mind To make a diagnosis one should feel the characteristic, unmistakable tumor If the baby has pyloric stenosis, the tumor can be felt in practically 100 per cent of cases In fact, our percentage for a short time was 101 per cent—we felt a tumor that wasn't there Standing at the left side of the baby, the examiner with his left hand palpates rather deeply in the right upper quadrant and slowly moves his hand upward and downward Suddenly it encounters a hard firm mass which feels like a peanut under a blanket After a bit of experience there is no mistaking the sensation imparted to the examiner's fingers The lower pole of the kidney, a transverse process of a lumbar vertebra or a transverse tendinous intersection of the rectus muscle is not easily mistaken for a pyloric tumor Either one feels the tumor or one doesn't When directing a student in the art of palpating a pyloric tumor, it is gratifying to observe the look of intense uncertainty suddenly give way to a look of victorious satisfaction as he announces, "Oh, now I feel it"

Babies don't like to have their abdomens palpated, so they cry, tighten their rectus muscles and make examination impossible To obtain cooperation, the baby is given a nipple generously coated with granulated sugar While he hungrily focuses his attention on this choice tidbit, his abdominal muscles relax and the examination may proceed Not infrequently it is impossible to feel the tumor at the first attempt If a tumor cannot be felt in a child whose history and symptoms are typical of pyloric stenosis, the baby, lying disrobed on a table, is given a 6-ounce bottle of formula and observed Peristaltic waves moving from left to right will be seen without fail if pyloric stenosis is present Rarely, one may feel a hitherto elusive tumor when the stomach is filled Usually, however, the best time to find the tumor is immediately after vomiting, during those few minutes while the baby is limp and the abdominal wall is completely relaxed

The roentgenologist facetiously says, "Why all this fuss about palpating the tumor? Why not send the baby to the x-ray department for a diagnosis? I will show you the diagnostic string sign in the pylorus" We stubbornly answer that we can make the diagnosis clinically and counter with the statement that we don't like to have barium in the stomach if immediate operation is being considered, but we do humbly admit at times that we need the help of the roentgenologist

with approximately 750 operations for pyloric stenosis performed during the past 20 plus years there has been but one death and that occurred in a three-and-a half pound six weeks premature infant upon whom I operated It has always seemed to me that I can detect on the part of the surgical staff when mortality is discussed a restrained but apparent satisfaction that the lone misfortune should have happened to the chief of the service

It is less than fifty years since Fredet Ramstedt and a host of unknown surgeons pediatricians physiologists chemists and others showed us the way to handle pyloric stenosis surgically The operation is simple and the results are excellent

infection We now make sure the stomach is empty Incidentally, the catheter is kept in the child's stomach during the entire operation

General anesthesia consisting of open drop ether is, we believe, the anesthesia of choice Local anesthesia is favored by some, but seems unwarranted for an operation which can much more easily be done under general anesthesia and in about half the time

A muscle-splitting incision is made high in the right upper quadrant The anterior sheath of the rectus is cut longitudinally, the fibers of the rectus muscle are spread with a hemostat, and the posterior sheath of the rectus and peritoneum are cut transversely The stomach is easily lifted from the abdomen with a forceps and the pyloric tumor exposed and delivered into the wound While holding the pyloric tumor between the thumb and forefinger of the left hand, a shallow cut is made through the serosa and just into the tumor in an area most free of blood vessels With a curved hemostat or some instrument modified to suit the pecadilloes of the surgeon the cartilage-hard fibers are split by tearing and separated until the layer of mucosal membrane bulges up between the split fibers

The only point at which trouble is encountered is the duodenal end of the tumor too vigorous separation will cause a tear in the duodenal mucosa, too timorous separation will fail to relieve the obstruction Every surgeon of considerable experience with pyloromyotomy has upon one or more occasions made a tear in the duodenum The tragedy in this operation is not that a hole is made in the duodenum, but that it is overlooked and not repaired An opening in the duodenum is recognized by the appearance of typical pink mucosa or a bit of stringy mucus in the lower end of the pyloric wound If there is any uncertainty about having made a hole in the duodenum, the abdomen should be filled with sterile salt solution Pressure on the stomach will force air through the opening in the duodenum, and bubbles seen rising through the water will leave no question A couple of well placed 5-0 silk sutures on a swaged-on needle are adequate to close the opening The filmy omentum is drawn over the suture line and fixed with another stitch

Four to six hours after operation the infant is started on the standard pyloric regimen of feeding A dram of water is given One hour later a dram of 1 10 dilution of evaporated milk in water is given At the next hour 2 drams of water are given and followed an hour later by 2 drams of the weakened formula The feedings are increased a dram at a time Usually, by the fifth to seventh day following operation, the child is beginning to regain lost weight and is ready for discharge from the hospital

RESULTS. Mortality following pyloromyotomy dropped precipitously during the twenties and thirties and during the past fifteen to twenty years has levelled off at a fraction of 1 per cent In our experience

Meckel's Diverticulum

Meckel's diverticulum is a loose term improperly applied to all remnants of the omphalomesenteric duct. This duct conveys nourishment from yolk sac to embryo in the oviparous Useless to the human embryo it normally disappears in early fetal life and leaves no remnants Failure of involution of the entire duct results in formation of a fistula extending from small intestine to umbilicus If only the umbilical portion persists a mucus-discharging sinus remains at the umbilicus Persistence of the intestinal end of the duct produces the most commonly found anomaly properly known as Meckel's diverticulum.

In view of the wide communication of yolk sac with primitive gut in the first few weeks of embryonic life it is not difficult to understand why islands of gastric and duodenal mucosa and even bits of pancreatic tissue are sometimes found in the diverticulum A diverticulum lined with normal ileal mucus membrane is a harmless deformity

intestinal juices and produce ulceration Microscopic studies will almost invariably show gastric mucosa in a Meckel s diverticulum which bleeds

Other complications of Meckel s diverticulum are perforation of a peptic ulcer and peritonitis Inflammation of the diverticulum may in all respects resemble acute appendicitis A fibrous cord extending from the diverticulum to some other structure or an unobliterated omphalo-mesenteric artery may produce mechanical intestinal obstruction

Before a definitive diagnosis is made of bleeding Meckel s diverticulum it is well to clear the large bowel thoroughly and look for a polyp in the rectosigmoid with a proctoscope and study the entire bowel roentgenographically with air-contrast medium in search for a polyp or polyps in the remainder of the large bowel Bleeding from a polyp is rarely so severe as from a diverticulum and the blood is apt to be bright red rather than brick red Nevertheless careful examination for polyps has at times been rewarding

Treatment

Surgical removal of a typical Meckel s diverticulum presents few problems If the diverticulum is long and there is no disease at its base a toothed ductus clamp is applied transversely the diverticulum amputated and the stump closed with a running suture of surgical gut for the mucosa and a row of interrupted silk sutures for the muscularis and serosa. If during the course of any routine abdominal operation an unsuspected Meckel s diverticulum is found, it is customary to resect it because there may be lodged in its tip some gastric mucosa capable of later causing trouble.

A Meckel s diverticulum may be missed at operation. A four year-old boy had had a couple of episodes of brisk rectal bleeding. At operation at a hospital ordinarily referred to as "elsewhere" no diverticulum had been found. Bleeding persisted. At a second operation a diverticulum was found in the mesentery of the bowel covered with adhesions It looked and felt like a large lymph node. All diverticula arise on the antimesenteric border of the bowel. So did this one. Previous infection and consequent adhesions had firmly plastered it to the mesentery

If the diverticulum cannot be amputated without encroaching on the lumen of the bowel, resection of a segment of ileum and end to-end anastomosis are in order

Diagnosis

Persistence of various portions of the omphalomesenteric duct determines what symptoms and findings may arise

An umbilical sinus resulting from a remnant of the proximal portion of the duct is a rare finding commonly mistaken for an ordinary umbilical granuloma A granuloma is grayish-white, nodular, firm and wet, whereas an umbilical sinus is soft, red and glistening and secretes mucus It looks like a small strawberry in the umbilicus A few applications of silver nitrate will eliminate a granuloma, but will have no curative effect upon a sinus Mucoid secretion from a sinus is sticky and, when touched, will make a string as the finger is withdrawn (The residents irreverently refer to this diagnostic procedure as "Potts' slimy test") Injections of Lipiodol into the sinus followed by roentgenograms will demonstrate a blind end in the abdominal wall or a direct communication with the bowel Even though the tract is open to the small intestine, there will rarely be a history of discharge of intestinal fluid or gas

Sudden, massive hemorrhage from the bowel in a child, usually between the ages of one and four years, is so characteristic of a bleeding Meckel's diverticulum as to be almost pathognomonic The condition is rare in infants below three months of age and in teen-agers Without premonitory or accompanying symptoms, the child suddenly passes a large amount of blood The blood is brick red What constitutes a large amount of blood is a matter of opinion Parents see some red discoloration in the toilet bowl and consider it a severe hemorrhage One asks the mother whether the child passed a teaspoonful or a cupful of blood If she promptly answers, "Much nearer a cupful," a bleeding Meckel's diverticulum is strongly suspected Whenever possible, more specific information is obtained by seeing the stool A mother, rather anxious about her two-year-old son's health, had heard of a bleeding Meckel's diverticulum She reported a couple of hemorrhages, but had not saved the stools In view of the absence of symptoms of hemorrhage in the child, she was advised upon the next occasion to save and send us a suspicious stool Instructions were followed, and in great agitation she sent in a specimen It was large, it was brick red It consisted largely of improperly masticated beets—the only vegetable the child would eat Sudden unexplained signs of hemorrhage, such as pallor, weakness and sweating, may be due to a bleeding diverticulum, unsuspected because the well oriented and independent child attends to his own toilet habits and is not introspective about color, amount or frequency

Hemorrhage from a bleeding Meckel's diverticulum arises from a peptic ulcer in misplaced gastric mucosa Acid secretions from the gastric mucosa trapped in the diverticulum escape neutralization by alkaline

Meconium Ileus

Meconium ileus is a manifestation of congenital fibrocystic disease of the pancreas Because of inadequate tryptic enzymes in the pancreatic juice of the fetus, undigested, sticky meconium accumulates in the ileum where it causes obstruction Fibrocystic disease of the pancreas, more descriptively referred to as mucoviscidosis, is recognized as the cause of intractable pulmonary disease meconium ileus is not a separate entity but a severe manifestation of the same basic pancreatic abnormality Approximately 10 per cent of infants with mucoviscidosis have meconium ileus.

Diagnosis

Meconium ileus in the newborn infant is not difficult to diagnose after one has seen a single typical case. The almost infallible clue is this the baby s abdomen is distended *at birth* Palpation at once imparts to the examining fingers a sensation of doughy resistance Loops of bowel

about the diagnosis to this point, the characteristic findings remove all doubt. The mid portion of the ileum is distended with meconium thick and so black that it gives the bowel the appearance and feel of a blood sausage From the distended segment there is a gradual decrease in the caliber of the ileum distally until a few inches or more above the ileocecal valve it is about the diameter of a lead pencil The jejunum is also dilated, but by liquid material and air The entire colon is small and contains brownish gray hard masses consisting of tenacious mucus and probably bits of meconium.

Different methods of surgical procedure have been advised but in the typical case we have performed the operation first suggested by Hiatt. With a fine needle considerable salt solution is injected into the distended ileum to free the meconium from the mucosa. We have used weak peroxide for this loosening process with success but some anxiety that too rapidly released oxygen might rupture the bowel. Recently a visiting surgeon suggested the use of a detergent It sounds like a good idea. A bit of gentle kneading is necessary to free the meconium from the mucosa. Then a transverse incision is made in the distal portion of the distended ileum, and the meconium is removed by gently milking, washing and grasping long masses with a forceps We once took out in one piece a cast over 1 foot long. After cleaning out all the obstructing meconium the ileostomy is closed with a few silk sutures Four grains five might do better of pancreatic granules mixed in about 10 cc. of water are injected into the ileum, and some water containing pancreatic granules is also injected into the colon to free and digest the sticky masses

Since we have had rather good luck with this procedure, we have been loath to change our method. However in case viability of the distended segment of ileum is uncertain exteriorization, resection and construction of a double barrel ileostomy is indicated, Gross prefers this method for all routine cases A rather good idea has been suggested by Koop The dilated segment of ileum is resected, and the proximal end is anastomosed to the side of the small-caliber ileum a few inches from the open end. This open end is then brought out on the abdomen where it serves not only for decompression of the small bowel, but also for irrigation of the colon if necessary

Postoperative Care

Postoperative care of these infants is demanding. After gastric suction for forty-eight hours the tube is removed and glucose water in small amounts is given frequently A daily enema containing pancreatin is of value for softening and loosening the mucoid nubbins in the colon These enemas must be given by the resident, who sees to it that the fluid

which feel as though they had been stuffed with putty are identifiable. If antepartum rupture of the ileum (usually due to volvulus) has occurred shortly before birth, the consequent chemical peritonitis manifests itself by a reddish hue in the skin over the entire abdomen, and pitting edema of the abdominal wall

After the first or second day of life the infant with meconium ileus presents the classic symptoms of intestinal obstruction—green vomitus and tympanitic distention of the upper abdomen Bowel movements consist of small, round, pellet-like masses of mucoid material, slightly more colored than those passed by the infant with atresia of the bowel, but definitely not normal meconium

Roentgenograms of the abdomen often show, as pointed out by Neuhauser, *a mottled appearance in distended loops of bowel due most* likely to tiny, trapped air bubbles in the inspissated meconium White described a roentgenographic sign of value in identifying meconium ileus distended loops of bowel, but *no* air-fluid levels in the upright films as ordinarily seen in typical intestinal obstruction

Masses of calcium salts deposited at the rim of the peritoneal cavity or scattered throughout the abdomen strongly suggest that antepartum peritonitis occurred a considerable time before delivery

Treatment

The choice of medical or surgical management will be determined by the severity of symptoms Personally, I have not seen a case of meconium ileus so diagnosed which could be handled medically, but from time to time about the country doctors tell of cases which have been successfully treated with persistent enemas containing pancreatic granules Infants referred to our hospital have required active surgical care

Over and over again it is repeated that the stomach must be emptied before every operation for intestinal obstruction and that a needle or tube must be inserted into a vein for administration of blood and fluid Meconium ileus is no exception

Under open-mask ether anesthesia a right paramedian incision is made Somewhat different findings are encountered in almost every case of meconium ileus Multiple fibrous adhesions, a sign of earlier peritonitis, may be found between loops of bowel through the entire abdominal cavity Fibrinous adhesions and cloudy fluid suggest recent bowel perforation Upon occasions volvulus with or without gangrene is seen In one instance in which the entire small bowel was gangrenous there was nothing to do but close the abdomen and inform the parents of the hopeless status

In the majority of cases, however, the greatly distended mid-ileum presents itself as the abdomen is opened If there was any uncertainty

Intussusception

When a mother anxiously telephones the pediatrician and says "This morning at twenty minutes past eight my baby had a queer spell. He pulled up his legs and screamed as though in terrible pain Then he got pale and broke out in a sweat. Now he just lies limp in his crib I can t understand it. He hasn t had a sick day in his life" the pediatrician recognizing the early cardinal symptoms of intussusception interrupts the mother s story and says "I'll be right over"

Intussusception is a strange malady of infancy It appears most commonly about age six months, but may be seen in newborn infants and occasionally in children five years old. About 80 per cent of intussusceptions occur during the first year of life. For some unknown reason, boys are more commonly affected than girls Stranger still is the fact that healthy bouncing infants are most commonly affected.

actually goes up into the large bowel Nursing regulations forbid nurses to give "high" enemas

As soon as it is established that the bowel has regained its tone and is functioning, feedings of weak, evaporated milk formula are cautiously begun If no vomiting occurs, 0 5 cc of Viokase is added to each bottle

After the child's discharge from the hospital the mother is instructed to continue the use of Viokase without fail and to increase the dose to 1 cc when the child is placed on solid foods Mothers sometimes get curious about the need for continuous medication and stop the Viokase, but the pungent odor of the stools the following day promptly stimulates them to renew the medication The child will have to take Viokase all his life

Medical supervision is constantly essential for these children Prophylactic doses of broad-spectrum antibiotics will be necessary indefinitely to prevent pulmonary infections It appears that the child who has recovered from meconium ileus is less subject to later pulmonary infections than the child who has the usual fibrocystic disease syndrome seen in pediatric practice

We fret and strain to save the precarious lives of infants jeopardized by meconium ileus Some die because of unavoidable, antepartum, intra-abdominal catastrophes, some die of complications following operation, and some die of uncontrollable pulmonary infections early in life The rest, between 50 and 60 per cent, thrive and seem quite normal Since recoveries from meconium ileus date back only a decade, predictions about the future are impossible It is doubtful whether any of these children will have anywhere near a normal life expectancy, but they have been given their right to live

finger is withdrawn Occasionally the intussusception resembling the feel of a cervix will be felt in the rectal ampulla

It is well to bear in mind that not all intussusceptions occur in healthy infants between four and five months of age. Within six months we saw two unusual cases in infants during the neonatal period. They were very ill with what was presumed to be bloody diarrhea. One was admitted to the hospital with this diagnosis the other I am ashamed to say was treated for this condition by us In neither infant could a mass be felt. Vomiting of bile and signs of intestinal obstruction guided us to the correct diagnosis but too late. Both infants died shortly after resection of irreducible intussusceptions

Children from three to six years of age also may have typical intussusception Elusive findings in a six year-old boy led to heated arguments and small wagers among the residents Some unquestionably felt a mass others were equally convinced that none was present. As is often the case both factions were right the intussusception causing minimal symptoms appeared and for no known reason spontaneously released itself During observation for a few days everyone eventually felt the "phantom mass." At operation nothing atypical was found.

Treatment

At the Children s Memorial Hospital the treatment of intussusception is surgical We have been accused of stubbornness by those who prefer attempted reduction by enema. As a matter of fact, protagonists for surgical reduction have been lined up against those favoring reduction by enema for many decades Feeling the sting of criticism, we tried enema reduction for a brief period and had some success Then one day a typical intussusception was reduced by barium enema and the process observed fluoroscopically As the intussusception unfolded itself barium seen entering the terminal ileum "proved" that reduction was complete. The child was better for a few hours and then had recurrence of symptoms. Twelve hours later at operation an ileo-ileal intussusception was reduced with considerable difficulty

We favor surgical treatment for a number of reasons

Ileo-ileal intussusceptions are not uncommon The telescoped ileum moves onward with peristalsis and passes through the cecum. This second mass involving the cecum is easily reduced by enema but the ileal portion is usually not reducible. Furthermore, the remaining obstruction in the ileum is not readily detectable fluoroscopically

An intestinal polyp or a Meckel's diverticulum as a starting point of intussusception is not common, but in those few cases in which they are etiological it is gratifying at operation to remove them

Duration of symptoms as a guide to reducibility is far from infallible-

Diagnosis

Pain is the first and most specific symptom The baby screams, cries whimperingly or is suddenly restless and appears startled He draws up his knees, becomes blanched and collapses in a state of shock The onset of pain is so sudden that the mother often will recall that the baby became ill, not some time this morning, but at 8 20 o'clock After a restful night and an early usual feeding she reports that he suddenly cried out in a different way than he ever had before After a short time the attack wears off, but the baby appears exhausted In the course of a few minutes or as much as a half-hour another, similar attack follows Of course the onset may be any time of the day or night If the physician happens to call between attacks, the mother, although quite convinced that the attack was abnormal, may apologize for being needlessly anxious

Vomiting is apt to occur with the first attack of pain If the time between spasms is long, the infant may take a feeding without coaxing only to vomit it with the next recurrence of pain The longer the intussusception persists, the more severe the vomiting becomes until it is bile stained and, finally, ominously brown

No other condition signals so definite a lead towards a correct diagnosis as the facial expression of a baby with intussusception During a spasm a sudden startled look of anxiety and pain crosses the baby's face As the spasm recedes the infant relaxes, becomes indifferent to its surroundings and is limp and pale "A far-away look in the eye" was a favorite expression of Dr Brennemann in describing the facies of a child with intussusception The alternating look of anxiety and indifference gives way to one of severe illness as pathologic changes progress

A mixture of mucus and blood in the stool resembling "currant jelly"—a horrible insult to this pungent sweet—often appears after intussusception has progressed for a number of hours If the history of intussusception is typical, but the child has passed a considerable amount of clotted blood in the stool, one may enjoy a moment of clinical distinction by making a diagnosis (proved at operation) of a bleeding Meckel's diverticulum as the starting point of intussusception

Although routine physical examination is in order in case of suspected intussusception, attention is focused upon the abdomen It's no use trying to palpate an abdomen during a spasm—nothing will be felt but rigid muscles After the spasm has passed and the infant's abdomen has become accustomed to the lightly stroking hand, a sausage-shaped mass can usually be felt in the region of the ascending or transverse colon It will be tender Robust infants with fondness for food are easily tricked into relaxing their abdomens for easier palpation by inserting into their mouths a nipple well coated with granulated sugar Finally, a rectal examination is in order Bloody mucus will often be seen as the gloved

Malrotation of the Bowel

Malrotation of the bowel is an inaccurate phrase commonly used to describe a symptom complex arising from failure of fetal bowel to find its proper place in the abdominal cavity During early embryonic life the colon, spearheaded by the cecum, doesn t rotate wrongly in case of malrotation—it simply fails to complete its normal course The cecum may stop anywhere on its counterclockwise route through the abdomen but usually is arrested in the right upper quadrant. The duodenum, instead of moving to the left as it should, extends from the pylorus straight down into the peritoneal cavity A membrane, most likely representing a leaf of misguided mesentery forms and extends over the duodenum from the posterolateral abdominal wall to the arrested cecum or descending colon Practically all difficulties arising from malrotation of

an intussusception with a five-hour history of symptoms may be reduced with difficulty, and one with a two- or three-day history may be unfolded with the push of a finger

It has been said that a weakened bowel may rupture under pressure of an enema I doubt whether this will happen if the reduction is done under fluoroscopic control and average judgment is used in the amount of pressure exerted However, the possibility of this disaster exists More important is the fact that valuable time is wasted

A scar on the abdomen is not a great price to pay for the peace of mind afforded by absolute certainty that the intussusception has been reduced Incidentally, it is now routine in our surgical group at the same operation to remove the appendix—a not inconsequential dividend for the patient

The operation is performed under open-mask ether anesthesia A McBurney's incision is made in those cases in which the mass is confined largely to the ascending or right half of the transverse colon With a finger the intussusception is easily milked back to the cecum With a thumb and forefinger the last portion is squeezed back into the cecum Attempts at reduction by pulling on the bowel will meet with failure The appendix is often black from interference with blood supply A right rectus incision is made in those cases in which the intussusception has advanced into the descending colon or protrudes from the rectum and in those patients whose symptoms suggest that resection may be necessary

Mortality following operation used to be rather formidable, but that has changed since the importance of blood replacement has become recognized Undoubtedly, many of the deaths following surgical reduction were due to shock caused by unappreciated blood and plasma loss During the past 12 years we have operated upon approximately 140 children with intussusception, with 3 deaths Two deaths were those in infants referred to above, and one death occurred in a child with extensive gangrene of the bowel necessitating wide resection and anastomosis

No! for the present we are not going to depend upon enemas for reduction, but shall continue to operate and go home to peaceful sleep

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the bowel are due, not to the fact that the bowel has failed to complete its intra-abdominal route, but to the abnormal membrane which is stretched across the duodenum. Ladd first pointed out the importance of this obstructing band. Another abnormality which occurs in conjunction with incomplete rotation of the bowel is failure of the mesentery of the embryonic mid-gut properly to attach itself to the posterior abdominal wall. The freely mobile loops of small intestine consequently are apt to become twisted.

Malrotation of the bowel is not difficult to understand if the complicated drawings of normal rotation of the bowel pictured in the anatomy books are momentarily disregarded. Chaffin and Snyder introduced an analogy to clarify the process. When driving an automobile, one properly grasps the steering wheel with one hand on each side, the left hand represents the duodenum, the right hand, the colon. Now make a left turn, the left hand (duodenum) moves to the left, and the right hand (colon) moves to the right. If the wheel is turned through an arc of approximately 180 degrees, rotation is complete, if the turn is 90 degrees, slightly more or less, rotation is incomplete, and the condition known as malrotation of the bowel exists. The process of development of a fibrous band across the duodenum, and improper arrangement and fusion of the mesenteric folds is poorly understood except by expert anatomists, and may be represented in this analogy only by false moves associated with making a left turn.

Diagnosis

The infant who vomits bile during the neonatal period, but has passed some normal meconium, most likely has incomplete obstruction of the duodenum, and the cause is usually malrotation of the bowel. The degree of duodenal obstruction determines the severity of symptoms. Persistent vomiting of all feedings and some bile the first day of life leads to a relatively early diagnosis. More confusing are those cases in which the infant vomits intermittently, for a few days all feedings are retained, then persistent vomiting follows for a day or two. This intermittent vomiting may go on for a few weeks, during which time the formula is changed, thickened feedings are tried, small doses of atropine are given, and repeated examinations are made for a pyloric tumor. Peristaltic waves may be seen passing across the stomach. The baby's bowels move scantily, weight is stationary, and the baby remains just to the side of adequate hydration. Then one day the physician sees *green* vomitus, recalls its importance, orders a roentgenogram and makes a diagnosis of incomplete duodenal obstruction.

Associated at times with symptoms of partial duodenal obstruction are ominous signs of strangulating obstruction due to volvulus of the

inadequately fixed small intestine The baby may pass blood in the stool or vomit dark blood because of venous engorgement of the obstructed circulation A mass representing loops of partially or completely strangulated bowel may be felt in the upper mid abdomen Signs of prostration are evident.

An infant with malrotation of the bowel may have such mild or intermittent duodenal obstruction that the true cause of persistent or periodic vomiting is overlooked for months or even years Such an infant eats poorly some days vomits sporadically a bit more than average and, in general, is a poor feeder During childhood he picks at his food and for no apparent reason has bouts of vomiting lasting a day or two Physical examination brings to light no abnormality Our staff saw in the out patient department a female infant who because of periodic vomiting was shifted from one formula to another She gained weight normally in spite of vomiting As she grew older a diagnosis of celiac disease was made and an appropriate diet prescribed By about two years as vomiting became periodic every month or two the diagnosis of celiac disease wore thin and was replaced by "cyclic vomiting" At age six years the child, not too bright, but well developed physically was labelled "neurotic and uncooperative" An unusually severe attack of vomiting finally occurred and required hospitalization Roentgenographic studies showing a dilated duodenum and the cecum in the right upper quadrant established the correct diagnosis Surgical release of the band causing duodenal obstruction not only stopped the vomiting, but also corrected the neurosis and lack of cooperation.

A history of vomiting bile or of excessive vomiting is always sufficient indication for an anteroposterior roentgenogram of the abdomen taken in the upright position. A dilated stomach, a bubble of air in the duodenum and shadows of air in the gastrointestinal tract immediately make a diagnosis of incomplete duodenal obstruction. In fact, a bubble of air identifiable in the duodenum is alone enough for a diagnosis because air is not normally seen in the duodenum A swallow of barium will confirm the findings of a dilated duodenum. A barium enema showing the cecum in the right upper quadrant although unnecessary as a guide to treatment, establishes a diagnosis of malrotation of the bowel

Treatment

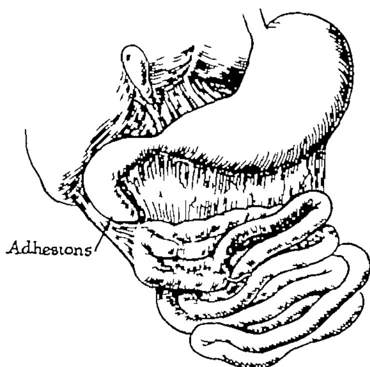
After such preoperative care as is necessary surgical exploration under open-mask ether anesthesia is in order The stomach, of course has been emptied. The abdomen is opened through a high, right paramedian incision and distended loops of bowel are allowed to herniate out of the wound Volvulus is looked for first and if severe enough to constrict the mesenteric vessels will immediately be recognized by the

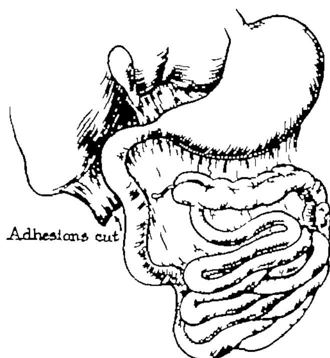
changed color of the bowel wall Normal-appearing bowel does not rule out the necessity of looking for volvulus unassociated with strangulation The bowel is untwisted in a counterclockwise direction and dealt with as its condition demands

The congenital band—a thin, translucent, bloodless, fibrous structure—obstructing the duodenum is sought for and identified After it has been cut transversely the cecum or transverse colon to which it is attached is loosened and can easily be elevated The duodenum, extending straight downward, is freed from any remaining obstruction At this point it is important to pass a no 16 French catheter through the mouth into the stomach, from where it is directed through the duodenum well into the jejunum This maneuver is essential to demonstrate patency of the duodenum In three instances we found an intrinsic partial obstruction of the duodenum not visible or identifiable by external examination The second obstruction was relieved by opening the duodenum and excising the diaphragm Because many of these infants vomit after operation it is reassuring to have demonstrated that the duodenum is widely open

The operation is finished No attempt is made to place the cecum in its normal position In fact, after the fibrous band has been cut the right colon tends to move a bit to the left If the child's condition is good and the operation has been short, the misplaced appendix is removed, if it is not removed, the parents are told that the appendix is in an unusual position and that if at any time signs of appendicitis arise, they so inform the surgeon

After operation a suction tube is left in the stomach for forty-eight hours, then removed, and feedings are begun Postoperative vomiting is sometimes persistent, necessitating prolonged intermittent stomach aspiration and intravenous administration of fluid, blood and plasma A three-weeks-old infant was operated upon for malrotation of the bowel At operation a catheter demonstrated duodenal patency, but the infant continued to vomit all feedings for about a week The abdomen was reopened, and a small catheter was again inserted through the stomach into the jejunum and left there It promptly backed up into the stomach, and for another week all feedings were vomited At the third operation a plastic tube, to the end of which was tied a short section of fine Penrose tubing, containing 0 25 cc of mercury, was introduced into the stomach through a nostril As it was impossible to direct this tube through the pylorus by external manipulation, a small transverse incision was made in the stomach, and with a hemostat the tube was pushed into the duodenum, from where it could be milked well into the jejunum As soon as the infant had recovered from the effects of operation, feedings by tube were begun and well tolerated The tube was left in the jejunum for two weeks without mishap or apparent irritation of the nose or throat


Adhesions


Adhesions cut

At the end of this time, while the tube was still in the jejunum, feedings by mouth were given for two days to be sure that the duodenum was open and functioning. No vomiting occurred, and the tube was removed. The child continued to gain weight and thrive. It may be assumed that the feeding tube acting as a support for the duodenum prevented angulation and recurrent mechanical obstruction. The residents jocularly referred to the tube as "Potts' infallible feeder." It worked.

During the period of vomiting and repeated operations this baby dropped in weight from 8 to 5 pounds and just before forced feedings were begun looked like a picked chicken. At age six months he had overcome the handicap, was physically up to standard, and psychically well adjusted. Apparently the trauma of repeated operations, innumerable needle pricks, nasal tubes and such distress as a sick baby suffers had left no scar on the miniature psyche or cells.

Atresia and Stenosis of the Intestine

INTESTINAL ATRESIA

Congenital atresia of the intestine is a relatively uncommon anomaly of the newborn An active obstetrical service in a general hospital will average less than two cases a year Because the condition is rare, early symptoms are apt to be overlooked until unmistakable signs of advanced intestinal obstruction manifest themselves.

Diagnosis

The first and most characteristic symptom of atresia of the bowel is vomiting, the most important feature of the vomitus is its color—green A

child with atresia of the duodenum may vomit bile within the first twelve hours of life The further distal the obstruction, the later the appearance of green vomitus However, within forty-eight hours after birth almost every infant with atresia of any portion of the bowel will have vomited some bile The fact that the baby has vomited bile is often not learned by the attending physician because his attention is not called to the telltale color The nurse simply records in her notes that the baby vomited The physician, if he looks at the nurses' record, sees a notation about vomiting, but because most infants frequently vomit or regurgitate, pays little attention to the observation We use an ungrammatical but expressive question when inquiring about the vomitus in any infant suspected of having intestinal obstruction "Does the baby vomit green?" *An infant who "vomits green" has intestinal obstruction until proved* otherwise If nurses in general realized the importance of their observations and reported them to the attending physician, earlier diagnosis would be the rule

The characteristic of the infant's stools comes up for similar consideration The newborn child, now a person, is moved to the nursery, where he is accorded the dignity of a separate clinical record On the pages in the nurses' notes is a column headed "stools" Anything expelled from the rectum is a stool, accordingly, each such occurreence is simply recorded by the nurse with a check mark under the proper heading The bowel movement may be a tiny mass of grayish-white, sticky mucus no larger than a lima bean, so characteristic of atresia of the bowel, but it receives on the chart the same check mark as the large mass of shiny, green-black sticky meconium passed by every infant with a normal gastrointestinal tract To get the nurses to describe the amount, color and consistency of all stools would be extremely helpful, to get those same nurses to save for the doctor's inspection every bowel movement of any infant suspected of intestinal obstruction would be utopian (A nurse accurately described in detail the stool of a child with suspected fibrocystic disease and added to her notes "Odor—terrific" A letter of commendation for this student nurse was sent to the nursing office for her significant and astute observation)

Examination of the abdomen of a child with duodenal atresia is misleading except to those who previously have stubbed their diagnostic toes The baby has vomited bile, but the significant finding of intestinal obstruction—a distended abdomen—is absent In fact, the abdomen is scaphoid for the simple reason that no air or fluid has passed into the intestine Close observation may reveal distention of the stomach in the left upper quadrant In contrast, atresia of the jejunum, ileum or colon is attended with early and noticeable distention of the abdomen Intestinal patterning is usually visible in proper light and almost always palpable by lightly stroking the fingers up and down over the abdomen

Rectal examination in an infant is apt to be misinterpreted It is

relatively easy to insert one's little finger—often the index finger—into the rectum of a child with atresia of the bowel The finger can be inserted about an inch when the sensation of obstruction is felt. Not infrequently one of the house staff will report that the infant has high atresia of the rectum. Actually what gives the impression of complete obstruction is the tiny bowel just above the rectum which will not admit the tip of a little finger Even in a child with a normal gastrointestinal tract the examining finger often seems to strike an obstruction in the region of the lower end of the rectosigmoid in the hollow of the sacrum

Roentgenographic studies are essential to confirm the diagnosis of intestinal obstruction As in all emergency conditions of the newborn demanding roentgenograms, it is essential to send the infant to the x ray department for clear pictures both in the erect and in the supine position It is doubtful whether x ray films taken in the supine position add any information, but roentgenologists request both views and we meekly conform

Absolutely diagnostic of complete obstruction of the duodenum is the "double bubble" shown in the x ray film The large air bubble on the left side is the dilated stomach the smaller bubble slightly to the right of the midline in the upper part of the abdomen represents air in the obstructed and dilated duodenum. No gas pattern will be seen beyond the duodenum.

Atresia of the jejunum is recognized roentgenographically by air fluid levels in a few loops of bowel, and atresia of the ileum by similar findings throughout many loops of the small intestine Tremendous dilatation of the easily distensible cecum makes a diagnosis of atresia of the colon.

To give barium by mouth to infants with suspected atresia of the duodenum or bowel is not only unnecessary for diagnosis but is also unwise because of the danger of vomiting and aspiration. Furthermore, after operation barium in the bowel is apt to cause obstruction at the anastomotic site There is some difference of opinion about the desirability of giving these infants barium enemas we rarely do because it is difficult to force the barium through the narrow undeveloped colon and little is learned from the procedure. The possibility of having barium plug up the large bowel and later interfere with bowel evacuation more than offsets the satisfaction of seeing a film of a well formed colon filled with barium. If there is any uncertainty about a differential diagnosis between Hirschsprung's disease and atresia of the bowel a barium enema is definitely in order (See Hirschsprung's Disease, p 195)

Treatment

The treatment of congenital atresia of the gastrointestinal tract is surgical. Before the infant is rushed to the operating room it is well to

talk plainly with the father and the grandmother, who likely have accompanied the infant to the hospital If it is obvious, for example, that the infant with duodenal atresia is a mongolian idiot—about 30 per cent are—a few words, gently spoken, about the possibility that the child may have other serious deformities, especially of the brain, are in order It is also explained that operative correction of these defects is hazardous and that in certain circumstances it is impossible to do anything for the child Nothing establishes better rapport between parents and surgeon than some kindly explanatory conversation about what is to be done and why Parents like to be made to feel that they are intelligent enough to understand a bit of explanation about the unfamiliar disease from which their child is suffering

Need for preoperative preparation with fluid and blood will be determined by the condition of the child Before the infant is anesthetized and as it is lying on the operating table, a no 16 or 18 French catheter is inserted into the stomach through the mouth, and all fluid is removed by suction with an Asepto syringe It is amazing how much liquid may be removed from the stomach even though the infant has never had a drink of water or a drop of formula Open drop ether is the anesthesia of choice A polyethylene tube must be placed in the saphenous vein not only for a blood transfusion during operation, but also for maintaining fluid and electrolyte balance afterwards

Surgical treatment for duodenal atresia is a side-to-side, retrocolic, isoperistaltic duodeno-jejunostomy performed with a continuous serosal suture of 5-0 heart silk for the serosa and 5-0 surgical gut on a swaged-on needle for the mucosa The anastomosis is performed in a routine manner, but, because the jejunum is of small caliber, special attention is given to placement of the mucosal sutures not more than 2 mm from the edge to avoid obstruction of the anastomotic channel at the lower angle When the anastomosis is completed, it is wise to demonstrate by pressure on the duodenum that air and fluid will go through the new opening

When the diagnosis is atresia of the small bowel, a right paramedian incision is made and the distended bowels are allowed to herniate out of the wound, where they are covered with wet lap pads Attempts to keep most of the distended bowel in the abdomen lead only to frustration and excessive trauma Upon exploration one most commonly finds distended bowel ending blindly, and, distal to this point, small-caliber bowel not more than 6 to 8 mm in diameter

Upon three occasions when opening the abdomen for congenital atresia of the bowel, we found throughout the peritoneal cavity extensive fibrous adhesions At the point of atresia in each instance lay a mass of semi-necrotic material Microscopic examination of this grossly amorphous tissue in one instance showed remnants of small bowel mucosa Presumably a loop of bowel had become twisted, ruptured and allowed

enough spillage to produce antepartum chemical peritonitis The ends of the bowel had sealed themselves and produced the typical clinical picture of atresia.

After identifying the point of obstruction sterile salt solution is injected with a fine needle into the small-caliber bowel to enlarge its diameter and to demonstrate patency of the bowel to the rectum Injection of some air with the salt solution produces bubbles which are visible through the bowel wall and can be identified as they pass distally Demonstration of patency to the rectosigmoid is important because at times there is a second obstruction not visible upon external examination

The distended bowel may be deflated by inserting a large needle at its very end, but usually it is necessary also to remove accumulated meconium with a suction tube For some years we performed a side-to-side anastomosis between the disproportionate sized segments of bowel but recurring obstruction followed so frequently that we turned to end to-end anastomosis We gave up this technique also because of too frequent leaks We now resect the greatly dilated distal segment—as many inches as are necessary to get to good healthy looking intestine—and do an end to-side anastomosis The dilated end is anastomosed to the side of the narrow bowel as near its proximal end as possible We use a continuous suture of 5-0 heart silk on the serosa and 5-0 catgut on a swaged-on needle for the mucosa. Again as stated above, it is important to place the mucosal suture near the edge to avoid obstruction at the distal angle of the anastomosis

In those unfortunate cases in which multiple atresias are found all one can do is to make a series of anastomoses knowing full well that the outcome probably will be fatal.

The abdomen is closed without drainage A plastic suction tube is inserted through the nose into the stomach, and the infant is returned to his oxygenated Isolette

Antibiotics are routinely prescribed. Physiologic saline solution 5 per cent glucose plasma and blood are given as needed. Suction is maintained on the drainage tube in the stomach for forty-eight hours Then it is removed, and thin feedings of 5 to 10 cc. of a mixture of 1 part evaporated milk and 10 parts of water are given each hour If the baby doesn t vomit within twelve to eighteen hours and begins sucking his fist, feedings are slowly increased until a maintenance diet is reached by about the fourth or fifth day after operation. If after feedings are begun vomiting recurs the stomach tube is reinserted and suction maintained for twenty four hours when feedings are again cautiously begun

Observation of every stool is in order when one sees some beautiful golden-yellow material as evidence that food has passed through the gastrointestinal tract, then one knows that the operation has most likely been a success.

talk plainly with the father and the grandmother, who likely have accompanied the infant to the hospital. If it is obvious, for example, that the infant with duodenal atresia is a mongolian idiot—about 30 per cent are—a few words, gently spoken, about the possibility that the child may have other serious deformities, especially of the brain, are in order. It is also explained that operative correction of these defects is hazardous and that in certain circumstances it is impossible to do anything for the child. Nothing establishes better rapport between parents and surgeon than some kindly explanatory conversation about what is to be done and why. Parents like to be made to feel that they are intelligent enough to understand a bit of explanation about the unfamiliar disease from which their child is suffering.

Need for preoperative preparation with fluid and blood will be determined by the condition of the child. Before the infant is anesthetized and as it is lying on the operating table, a no. 16 or 18 French catheter is inserted into the stomach through the mouth, and all fluid is removed by suction with an Asepto syringe. It is amazing how much liquid may be removed from the stomach even though the infant has never had a drink of water or a drop of formula. Open drop ether is the anesthesia of choice. A polyethylene tube must be placed in the saphenous vein not only for a blood transfusion during operation, but also for maintaining fluid and electrolyte balance afterwards.

Surgical treatment for duodenal atresia is a side-to-side, retrocolic, isoperistaltic duodeno-jejunostomy performed with a continuous serosal suture of 5-0 heart silk for the serosa and 5-0 surgical gut on a swaged-on needle for the mucosa. The anastomosis is performed in a routine manner, but, because the jejunum is of small caliber, special attention is given to placement of the mucosal sutures not more than 2 mm. from the edge to avoid obstruction of the anastomotic channel at the lower angle. When the anastomosis is completed, it is wise to demonstrate by pressure on the duodenum that air and fluid will go through the new opening.

When the diagnosis is atresia of the small bowel, a right paramedian incision is made and the distended bowels are allowed to herniate out of the wound, where they are covered with wet lap pads. Attempts to keep most of the distended bowel in the abdomen lead only to frustration and excessive trauma. Upon exploration one most commonly finds distended bowel ending blindly, and, distal to this point, small-caliber bowel not more than 6 to 8 mm. in diameter.

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Many perils attend the postoperative course of these infants, whose start in life has often been hampered by prematurity underweight, other congenital deformities and advanced symptoms of obstruction a leak in the anastomosis may produce local or generalized peritonitis obstruction may recur and demand a second operation an infected wound may cause dehiscence and pulmonary complications may follow aspiration of vomitus. Diagnosis of atresia of the bowel is simple operative correction is routine postoperative care is demanding mortality in all cases fluctuates between 30 and 50 per cent

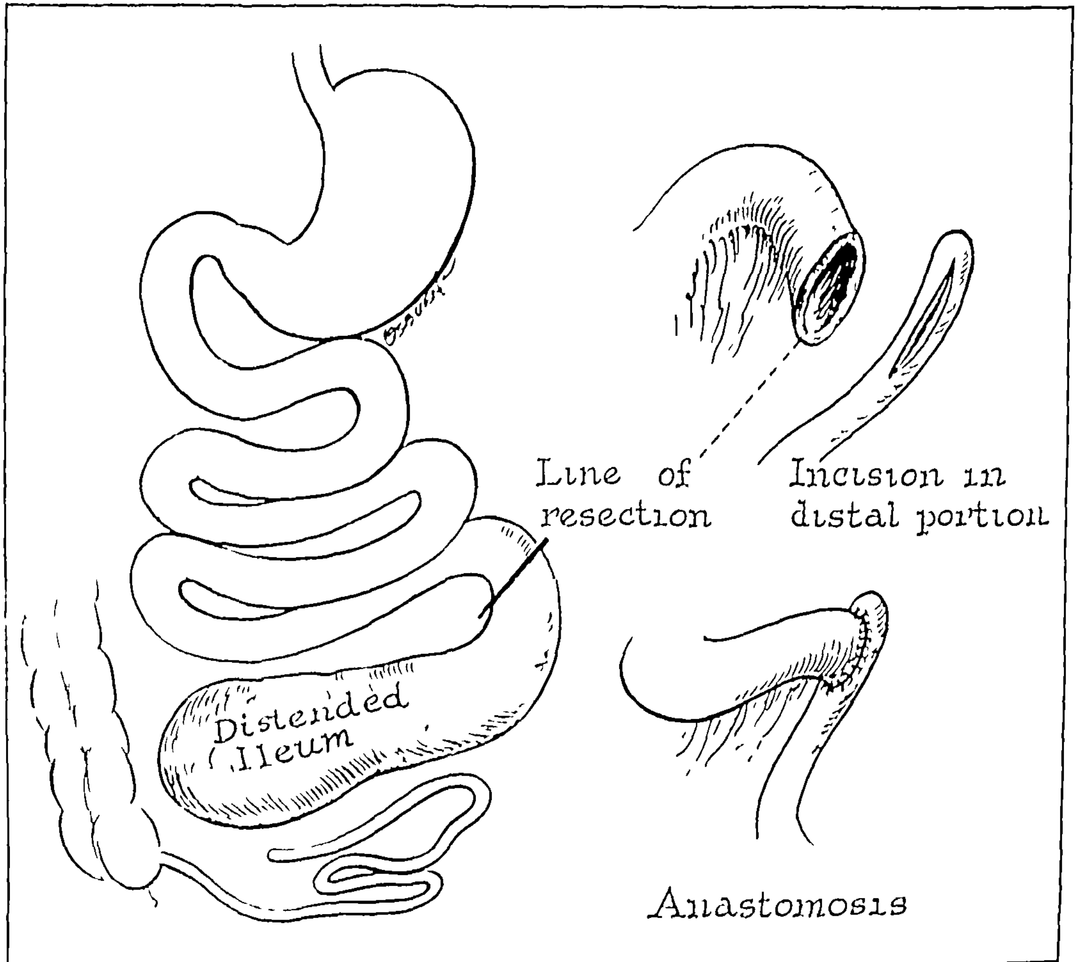
STENOSIS OF THE BOWEL

Congenital stenosis of the gastrointestinal tract in infants and children most commonly occurs in the duodenum Vomiting is the outstanding symptom and varies with the degree of obstruction Often the baby is tried on many different formulas without success In mild cases periodic vomiting which persists through early childhood is ascribed to allergy celiac disease "cyclic vomiting," nervousness psychic maladjustment and finally in desperation to "Oh some children just vomit easily" before the true cause is suspected. Physical and psychiatric examinations bring to light nothing more than an obviously underweight nervous irritable child. Prolonged observation of the abdomen after eating is occasionally rewarded by the appearance of telltale peristaltic waves moving from left to right. Roentgenographic examination with barium which demonstrates a dilated duodenum clinches the diagnosis of stenosis due to extrinsic pressure by congenital bands or an annular pancreas or to intrinsic malformations such as webs or fibrous annular bands

Surgical procedures will have to be adapted to the lesion found at operation For relief of duodenal obstruction caused by an annular pancreas a bypassing duodeno-jejunostomy will be safer than splitting the pancreas and thereby inviting the formation of a pancreatic fistula. Complete division of external obstructing congenital bands such as are associated with malrotation of the bowel will effect a cure

Intrinsic duodenal obstructions are best relieved by duodeno-jejunostomy if the stenosis is firm and hard and involves a considerable segment of the bowel wall Resection of an obstructed duodenum and end to-end anastomosis are rarely possible without imperilling important structures If by palpation a narrow soft obstruction is identifiable, it is acceptable to open the duodenum longitudinally and excise the weblike diaphragm The duodenum is closed directly or transversely depending upon the degree to which the obstruction has been relieved

Stenosis of the jejunum or ileum, unless severe is rarely diagnosed or suspected until a bolus of food or a foreign body becomes incarcerated in the constricted area and produces complete obstruction One child


Line of resection
Incision in distal portion
Distended Ileum
Anastomosis

Many perils attend the postoperative course of these infants whose start in life has often been hampered by prematurity, underweight, other congenital deformities and advanced symptoms of obstruction. A leak in the anastomosis may produce local or generalized peritonitis, obstruction may recur and demand a second operation, an infected wound may cause dehiscence, and pulmonary complications may follow aspiration of vomitus. Diagnosis of atresia of the bowel is simple, operative correction is routine, postoperative care is demanding, mortality in all cases fluctuates between 30 and 50 per cent.

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with diffuse abdominal complaints finally showed signs of intestinal obstruction. At operation a button was found jammed against the stenotic barrier. Resection of the involved segment and end-to-end anastomosis were followed by relief of all symptoms. Another infant had an altogether similar history, but because of his rather poor physical condition a by-passing jejuno-jejunostomy was performed. That was a mistake. Two years later the child returned with a history of abdominal distress, poor appetite, loss of weight, pallor, and signs of recurring obstruction. A large diffuse mass was palpable in the right upper quadrant. At operation a single, hugely distended loop of bowel was found—one end distal, the other end proximal to the previous side-to-side anastomosis. The dilated loop of bowel and the old anastomosis were resected, and an end-to-end anastomosis of the jejunum was performed. The child was cured.

Appendicitis and Appendiceal Abscess

ACUTE APPENDICITIS

Acute appendicitis probably not as common today as it was twenty five years ago receives less attention because the fear of killing complications of the disease has been largely eliminated by the advent of antibiotics At the turn of the century an accurate diagnosis of acute appendicitis followed by a successful appendectomy was an event worthy of prideful review even by professors of surgery Today the junior surgical resident removes an acutely inflamed appendix and nonchalantly reports to his superior "Just a routine appendectomy" It is now rare for a patient to die of acute appendicitis and its complications The surgeon has been freed from that helpless feeling of being able to do nothing but

depend upon the recuperative powers of the patient when confronted with a ruptured appendix and generalized peritonitis. Although the *terrors of complications of acute appendicitis have been allayed,* the importance of recognizing the disease in its early phases is as great as ever.

Actually, there is little to be said on this subject of acute appendicitis that has not been said a thousand times, and yet a book on surgical problems in children would not be complete without repeating a few of the cardinal points about this common ailment. I doubt whether there is any disease which has symptoms of such variable degree as appendicitis. Everyone has seen children with almost negligible symptoms who, at operation, have had gangrene of the appendix. It is these mild cases, so easily missed, which eventually make every clinician of experience eat his portion of fricasseed crow.

Appendicitis is rare during the first year of life and distinctly an oddity during the first six months. A few cases appear in the second year, but by the fourth to sixth year it is common.

Diagnosis

Pain is the first symptom of acute appendicitis. Parents are inclined to believe that the pain must be severe to warrant operation. At times pain is acute from the onset of disease, but more often it is nothing more than a generalized stomach ache hardly severe enough to keep the child from play or from his favorite television program. The pain is generalized and continuous for a number of hours before it settles down somewhere in the vicinity of the right lower quadrant. Intermittent, colicky pain is indicative of gastroenteritis—often preceding diarrhea, dietary indiscretions or simple constipation rather than appendicitis. Pain that truly begins in the right lower quadrant may be, but likely is not, that of appendicitis. Again one bears in mind that the phase of generalized stomach ache may have been mild and forgotten. Nausea and vomiting are so common to most diseases of childhood that they are not necessarily symptoms of acute appendicitis. However, when vomiting occurs *after* the onset of pain, it begins to take on significance. It is uncommon for a child *with acute appendicitis not to vomit.* In fact, if a child complains of persistent stomach ache for a number of hours and does not have either nausea or vomiting, the chances are good that he does not have appendicitis. It has been so rare in my experience to see a child with acute appendicitis who has not had nausea and/or vomiting that in their absence we prolong observation for a few hours. If the signs and symptoms of appendicitis are fairly typical, but the child has not vomited, we ask him how he would like a big hot roast pork sandwich with lots of gravy. If he brightens up and says, "Yes," operation is delayed. If he squirms at the thought of such food, we examine him again.

The majority of children have some fever with acute inflammation of the appendix To say that a child with pain in the abdomen does not have appendicitis because his temperature is normal is just as foolhardy as to say that a similar child with fever does have appendicitis A degree or two of fever is present in the typical case but the warning of one of my confreres is of value if the advice is not taken too literally He said that he never took the temperature in a case of suspected appendicitis because he was afraid he would be influenced by it rather than by the symptoms and physical findings A child may have a ruptured appendix and a rectal temperature of 98° F or a simple acute appendicitis with a temperature of 105° F

Leukocytosis like fever is caused by innumerable infections and is tremendously variable It too is worthy of consideration but broad interpretation A white cell count of 18 000 to 20 000 per cubic centimeter with minimal but characteristic symptoms of appendicitis is significant Typical symptoms and physical findings of acute appendicitis cannot be disregarded in the presence of a normal white cell count. When findings are typical, the white cell count is apt to be somewhere between 15 000 and 25 000

The doctor who sees a patient at home with a suspected diagnosis of appendicitis is practically forced to do a "blood count" because parents have a fixed idea that the diagnosis cannot be made without it. Demanding parents will not hesitate to ask, "But, doctor how can you say this is appendicitis when you haven t done a blood count?" Of course their question is appropriate because a febrile illness with symptoms mimicking appendicitis and a white cell count of 4000 per cubic centimeter just could be measles

Physical examination of a child with abdominal pain calls for patience and gentleness General inspection reveals much At once it is decided—this child is sick, or this child does not appear ill The sick child who has acute appendicitis is apt to hold the abdomen fixed and to flex the right thigh. In contrast to the child with tonsillitis or otitis media who moves about the bed freely the child with an acute intra abdominal infection lies quietly in bed and does not wish to move or to be moved A typical history plus an anxious look, sunken lusterless eyes, a dry mouth, parched lips and a dry skin makes a presumptive diagnosis of perforated appendix and probably peritonitis

On the other hand, one justifiably suspects that the child who bounces about on the bed, smiles and converses freely does not have appendicitis even though the history is typical

The abdomen should be inspected for distention and localized swelling The first touch of the abdomen should be a light and superficial sweep of the hand to avoid setting the abdominal muscles in spasm to gain an impression about the presence or absence of rigidity and to learn

where the tenderness is greatest Then deeper palpation is begun in an area far removed from the point of tenderness Slow deep pressure followed by a sudden release has no place in the examination of a child's abdomen If the findings are minimal, equally deep pressure in both lower quadrants simultaneously will bring out differences in tenderness on the two sides It requires only a moment to feel for the spleen and liver edge Impaction of feces in the left colon is easily detected

Localized tenderness is the most important diagnostic point in the diagnosis of acute appendicitis If I were to be limited to one finding, I should choose finger-point tenderness Often children's confidence is gained by asking them to point to the sore spot Even though they misguide you, this spot is noted and is slowly approached from a point where there is no tenderness It may be required that one start on the chest or thigh to convince the child that he is not going to be suddenly hurt Finally, as confidence is established, one gently approaches McBurney's point A vocal response, a wince or a cry will indicate when the sore place has been touched As a matter of fact, sufficient information may be gained from the feather touch of the abdomen that it will be unnecessary to hurt the child If the localized tenderness is identified surely and repeatedly over or near McBurney's point, the child may be assumed to have acute appendicitis until proved otherwise If the local tenderness is elsewhere than over McBurney's point, but the other findings are typical, the presumptive diagnosis is still acute appendicitis If the appendix is retrocecal, gentle but fairly firm and deep pressure may be needed to find the tender area An appendix inflamed only at the tip and hanging deep in the pelvis may reveal a point of local tenderness only on rectal examination In doubtful cases look for that area of finger-point tenderness and do not underestimate its value.

When spasm of the rectus muscle has developed, there usually is little question about the diagnosis of an "acute abdomen" By the time rigidity appears, if the tenderness is still greatest on the right side and the history is typical, the appendix is acutely inflamed, probably suppurative or even gangrenous The presence of rigidity, if at all intense, necessitates evaluation of the type and degree of infection and whether or not the infection is still localized Some rigidity is often present in acute appendicitis, but one is always happier to have seen the child and to have made a correct diagnosis of appendicitis before much rigidity has developed

A digital rectal examination should finally be done, not because it is necessary for a diagnosis of a typical case, but because as a matter of training it must never be neglected as part of an abdominal examination A rectal examination is distasteful to children They will fuss and say it hurts everywhere because they dislike the examination About all one gains from a rectal examination is the finding of an appendiceal mass or the tip of a low-lying inflamed appendix which is very tender when

touched If the rectum is loaded with hard feces and symptoms of appendicitis are minimal an enema may effect a cure

The typical case of acute appendicitis can be diagnosed correctly by telephone, but the atypical case defies the diagnostic acumen of the most skilled and rudely trips the overconfident It is doubtful whether there is a diagnostician of many years experience who has not at some time or another gone astray in the diagnosis of acute appendicitis To see a skilled pediatrician tricked by an acute appendicitis presenting bizarre symptoms is comforting to those of us who already belong to that group who have made such misjudgments These comments are not made to excuse an error in diagnosis but to emphasize that a conscientious diagnosis of appendicitis in a child is one of the most difficult we are called upon to make

Of all the children with "stomach ache" about one in fifty has acute appendicitis The younger the child the more difficult it is to pick out unerringly those who have appendicitis Even the most careful and keen pediatrician will eventually join that humbled group who have allowed an appendix to rupture right before their eyes This does not mean that one should promiscuously advise appendectomy for any suspicious pain in the abdomen. It does mean that the utmost in diagnostic skill must be used to keep errors at a minimum

Differential Diagnosis

Acute appendicitis can be mimicked by innumerable diseases The error of "missing an appendix" is probably not so great as mistaking some other disease for appendicitis To remove a normal appendix is irritating to the surgeon but not a serious mistake unless the operation aggravates the overlooked disease

A sore throat, tonsillitis otitis media and pneumonia may cause abdominal pain. No child, no matter how typical the findings should ever be subjected to appendectomy before the upper respiratory tract has been studied. High fever rapid respiration dilatation of the alae nasi and leukocytosis suggest *pneumonia* During the first day of illness physical and x ray examination of the chest may give normal findings I have seen left lower lobe pneumonia simulate acute appendicitis The abdomen of a child with pneumonia feels different from that of a child with a similar fever caused by appendicitis The muscles are as a rule less rigid, the tenderness is more generalized, and the abdomen has a sort of "doughy" feel.

Gastroenteritis especially during mild epidemics when associated with abdominal pain, vomiting and slight fever constantly worries the physician. It is true that diarrhea is rarely a symptom of appendicitis but a mother s interpretation of what constitutes diarrhea makes it necessary to see many of these children on suspicion Tenderness in gastro-

enteritis is not localized as in appendicitis On the other hand, constipation may simulate appendicitis

Acute mesenteric lymphadenitis so closely mimics acute appendicitis that differentiation is impossible If you make a diagnosis of acute mesenteric lymphadenitis, it is wise to advise appendectomy to see whether you are right, and probably to have an acutely inflamed appendix removed

Acute pyelitis may be associated with fever, abdominal pain and vomiting The order of appearance of symptoms is different than in acute appendicitis The attack is often ushered in with a chill and high fever Chills are practically never associated with appendicitis until the infection has spread beyond the appendix Pus cells in the urine suggest pyelitis, but one must still be wary because an infected retrocecal appendix lying over a ureter will not infrequently cause red and white cells to appear in the urine

Before the discovery of antibiotics it was important to differentiate primary peritonitis from acute appendicitis because operation on the former was harmful Today the fear of primary peritonitis is less acute, and if there is any question about the accuracy of the diagnosis, appendectomy is advised

There are many more conditions which must be kept in mind, not only when unusual symptoms simulating appendicitis present themselves, but also when the case seems to be classic An eagle eye must be kept open for a beginning rash, petechiae suggesting Henoch's purpura, conjunctivitis suggesting measles, and so on Listen for a pleural friction rub or a pericardial rub when the history of acute appendicitis is irregular and the abdominal findings are atypical Rheumatic fever or osteomyelitis of the ilium may simulate appendicitis Persistent vomiting may mean intestinal obstruction due to adhesions from a previous operation, congenital bands, volvulus or an incarcerated hernia

A serious error is to mistake for appendicitis the vomiting and abdominal distress that go with *acidosis* from diabetes or other causes Examination of the urine, always a routine procedure before operation, will reveal the cause A child may have acidosis from persistent vomiting associated with appendicitis, but it can easily be differentiated from that due to diabetes or kidney disease

Putting one's hand under the head and testing for a stiff neck will avoid the occasional disaster of mistaking *meningitis* for acute appendicitis

Treatment

The treatment of acute appendicitis is unequivocal—immediate appendectomy In the average case little preoperative treatment is necessary

If the state of hydration is poor the proper type of fluids may be admin istered preoperatively and continued after operation The child has almost invariably vomited preoperatively but if there is any question about the stomach being empty a stomach tube should be passed It is always dangerous to give a general anesthetic to a child who has food or fluid in the stomach

After a simple appendectomy little postoperative care is necessary Foods are allowed as desired The child is allowed out of bed as soon as it is apparent that no complications are arising The hospital stay need be only a few days when all goes well

In children the operation can be done most safely under open mask ether anesthesia, using a McBurney or muscle splitting incision

In spite of continuous propaganda to the public against disregarding "stomach aches" and trying to cure them with catharsis the ratio of acute to perforated appendicitis remains about the same in children in the Chicago area. A study of appendicitis in children over the 15-year period from 1921 to 1935 revealed 40 per cent perforated with consequent gen eralized or local peritonitis During recent years a study of acute appen dicitis in the same class of people brought out the fact that 41 per cent of appendixes were ruptured before admission.

PERFORATED APPENDIX The treatment of the perforated appendix with generalized peritonitis is surgical The patient s preoperative con- dition is more carefully evaluated than in case of a simple acute appen dicitis Electrolyte and acid base balance are studied and corrected if abnormal Obviously the operation cannot be delayed until the child is completely hydrated but a few hours may well be spent in preparing for operation on the severely ill and dehydrated child.

The operation should always be done through a McBurney incision The pus is aspirated and the appendix removed with a minimum of trauma to the adjacent bowels The question of drainage of the abdominal cavity is still not settled Those who practically never drain and those who almost invariably do both claim they are right and have statistics to prove it. As usual, a middle ground seems safer After having given allegiance to both camps it now is our policy in cases of doubt to lay a soft Penrose drain in the lateral abdominal gutter No harm has come from this drain and formation of secondary abscesses we believe has been avoided.

Infection and abscess of the abdominal wall are the most frequent complications following removal of a gangrenous or ruptured appendix. Intra abdominal collections of pus may occur anywhere but most com monly are pericecal, pelvic and subphrenic. The temperature curve is the best early guide to the recognition of abscess formation The rectal temperature will vary between 101 and 103 F each day The important feature of the curve is that the temperature does not drop to normal in

the morning The temperature of patients with adequately draining abscesses may rise during the day, but in the morning the temperature is subnormal

The very ill child with complications following a perforated appendix *requires the usual attention to fluid intake, proper nourishment* and maintenance of a fair protein level The virtues of blood and plasma transfusions are so well recognized that no comment is necessary During prolonged feeding with artificial foods one should not overlook the necessity of vitamin administration

A Levin tube is routinely inserted through a nostril into the stomach and connected with a suction apparatus This tube is kept in the stomach until auscultation of the abdomen reveals that peristalsis has been re-established *or until the patient passes flatus spontaneously* While the tube is in place the child is given sips of water which serve to keep the tube open and to improve his morale As soon as it is obvious that the dangers of peritonitis are past, a liquid diet followed by a soft diet is given If, as occasionally happens, signs of intestinal obstruction appear due to plastic exudate about loops of bowel, the Levin tube is reinserted Usually, in a few days, if the bowel is kept empty, the fibrinous plaques dissolve themselves and normal flow of intestinal content is re-established

The prognosis in acute appendicitis and its complications improved slowly with each decade until the advent of antibiotics, then it brightened miraculously From 1921 to 1925 the mortality rate at the Children's Memorial Hospital was 10 2 per cent for all cases and 21 per cent for perforated appendixes During recent years since all antibiotics have been freely available, the mortality rate has dropped to 1 4 per cent for all cases The antibiotics are truly a miracle of modern medicine

APPENDICEAL ABSCESS

The formation of a walled-off mass about an inflamed or ruptured appendix *is nature's successful method of limiting the spread of infection*

One anticipates finding an appendiceal abscess in a child who is brought in after the third or fourth day with a typical history of acute appendicitis On examination one occasionally sees and frequently feels a mass in the right lower quadrant or detects a pelvic mass on rectal examination

In determining the proper treatment of an appendiceal mass, it is essential to picture mentally in each case the pathologic changes occurring The omentum, as the abdominal lifesaver, wraps itself about the infected appendix and, if the rupture is small and the infection not too virulent, walls off the infection The mass felt in such cases contains little pus and consists largely *of an edematous roll of omentum* and a heavy plastic exudate In the more virulent forms of appendiceal abscess the

mass consists of a pyogenic membrane enclosing moderate to large amounts of pus

Before the introduction of antibiotics our attitude towards the treatment of appendiceal abscess was conservative—nonoperative except in large or growing abscesses Mortality statistics supported the soundness of this form of treatment Today our attitude is still conservative but we operate on more appendiceal abscesses in the early stages of their development than previously because of our faith in antibiotics to limit the spread of the infection we may have seeded during operation A child who enters the hospital with a mass in the abdomen and with a history of appendicitis is put under observation The duration of illness temperature size of the mass and general condition help us to decide on the course of treatment. If the clinical condition is good if the mass is well walled off and if there are no signs of intestinal obstruction, the child is given antibiotics and a soft or liquid diet In the course of a few days to a week the temperature becomes normal and the mass recedes Six weeks after the mass has disappeared an appendectomy is done It is essential that the appendix be removed within a couple of months after recovery because of its tendency to rupture a second time The paucity of adhesions found at operation after disappearance of an undrained appendiceal abscess is amazing.

In the days before antibiotics I am sure mortality statistics in appendiceal abscess were better in general with conservative than with operative treatment. Now it is established that the antibiotics play a great role in minimizing the battle of peritonitis Consequently more appendiceal abscesses are operated upon in the relatively early stages of development.

Pelvic abscesses which after a reasonable length of time do not drain spontaneously may be carefully opened through the rectum Usually they drain spontaneously

CHRONIC APPENDICITIS

No subject arouses more animated discussion than that of so-called chronic appendicitis This term includes all those appendixes associated with bizarre symptoms and indefinite distress in the region of the right lower quadrant It does not include those patients who have a definite attack of appendicitis which terminated in an appendiceal abscess or those who had a rather typical case of appendicitis, but for one reason or another were not operated upon Intermittent appendicitis is a proper term for such conditions and the treatment is appendectomy Chronic appendicitis has also been labeled mechanical appendicitis and more recently has been referred to as appendicopathy High sounding phrases do not change the picture of chronic appendicitis

My training included the oft-expressed axiom, "There is no such thing as chronic appendicitis" With this swallowed but undigested bit of learning I began the practice of surgery A mother called me soon thereafter to see her seven-year-old boy with her self-made diagnosis of appendicitis The child had been sent home from school because of "stomach ache" He was a skinny little fellow who ate poorly and often had twinges of abdominal pain on the right side His temperature was normal, his leukocyte count was 10,000 per cubic millimeter The minimal tenderness in the right lower quadrant was unconvincing, and on a bland diet he promptly recovered The mother was reassured that the child had nothing whatever wrong with the appendix Time went on and repeated minor episodes recurred, in one of which I saw him and again virtuously denied any lesion in the appendix In the course of months, one morning the boy was sent home from school with another attack Oddly enough, I was called again Examination still yielded the same indefinite findings as before His temperature by mouth was 99° F His leukocyte count was 9000 I saw doubt and dismay in the mother's eye, was distressed by the undigested bit of wisdom of student days, but advised appendectomy. The appendix, not inflamed, but containing a fecalith 2 cm long and 1 cm in diameter, was removed During the following year the patient gained 20 pounds in weight There was no recurrence of pain The child's irritable disposition was transformed, and my education on the subject of chronic appendicitis began Let no one conclude that indefinite and troublesome pains in the region of the abdomen can be cured by appendectomy

Before appendectomy for chronic appendicitis is advised, careful study of the child is necessary His poor adjustment at home or in school may be a factor in producing symptoms The stools should be examined for all types of infestation Constipation should be corrected Far more important than a fluoroscopic examination of the colon is a thorough study of the genitourinary tract for congenital anomalies, stones and infections Tuberculous infection anywhere in the gastrointestinal tract or lymph nodes may simulate chronic appendicitis After the child as well as the parents has been carefully studied and no cause for the right lower quadrant pain has been found, appendectomy may be advised with the definite understanding that a cure cannot be guaranteed Disappointments and successes will be about equally divided

Hirschsprung's Disease (Aganglionosis)

Fifty years before Swenson focused attention upon the distal spastic segment of large bowel so typical of Hirschsprung's disease, it was known that this segment was aganglionic. During all these years surgeons attacked the enlarged portion of colon, not realizing that dilatation was an effect rather than a cause. Although the last word by no means has been written about Hirschsprung's disease, it appears that resection of the spastic segment is physiologically sound

Diagnosis

Hirschsprung's disease in the newborn is easily recognized. Abdominal distention and vomiting within the first few days after birth are always indicative of intestinal obstruction. The distended infant who has passed some normally slimy, tenacious, greenish-black meconium—even as little as a teaspoonful—almost certainly has Hirschsprung's disease. Rectal examination of the distended infant is always in order. Normal meconium may follow withdrawal of the examining finger in a child who to that time has passed nothing. Infants with atresia of the bowel or meconium ileus do not pass normal meconium.

Air-fluid levels will be seen in x-ray films taken in the upright position. Additional support for a diagnosis of aganglionosis is obtained if distention in the large bowel can be recognized roentgenographically. It is difficult, however, to distinguish the shadows of large from small bowel obstruction in an infant. A barium enema is of value in establishing the diagnosis.

An infant may go through the neonatal period manifesting fluctuating signs of incomplete intestinal obstruction. One day the child vomits everything, the next retains all food. The infant is pale and listless and fails to gain weight. The abdomen is constantly tympanitic. Enemas are only partially successful in relieving distention. Frequently a vigorous rectal examination will be followed by an explosion of gas and feces. An infant presenting such a clinical picture has aganglionosis of the bowel.

Diagnosis in children between ages two and four years is easy in the typical case and difficult in the atypical.

The child who, since the day of birth, has rarely moved his bowels without an enema, has a distended abdomen loaded with fecal masses and gas, and is pale, apathetic and underweight, probably has aganglionosis of the distal colon. Roentgenographic demonstration by barium enema of a narrow distal segment of bowel and a greatly distended colon clinches the diagnosis. Considerable skill and experience are required of the roentgenologist, not only in technique of giving the barium enema, but also in the interpretation of x-ray films. A colon thoroughly cleared of all fecal impactions is essential for proper and authentic x-ray studies.

The child whose troubles with constipation began by about two years of age probably does not have Hirschsprung's disease. Careful probing of the history is important. Practically every child who has aganglionosis of the bowel has a history of some degree of constipation since birth.

At this point analysis of constipation and methods of toilet training are investigated. Some children atavistically hate toilet training, potties, soap sticks, enemas, "grunt for mommy," and all the other tricks that go with maneuvers of getting the child to conform to the ways of civiliza-

tion Often a new baby in the house adds to the negativistic attitude of the older child The adored infant is an interloper who has dethroned the king, and something has to be done about it The best way of regaining attention is to refuse to comply with mother s wishes Somehow the two- or three-year-old discovers that refusing to "go potty" gets the best results Some children plagued by the green-eyed monster jealousy become verbal stutterers others colonic stutterers In those families in which the battle for bowel movements is relentless real constipation with consequent megacolon is the result

It becomes the duty of the physician to differentiate the acquired megacolon from that due to improper innervation In case of acquired megacolon the rectum is always filled with feces right down to the external sphincter in case of aganglionosis the ampulla of the rectum is usually not filled although it may be Mechanical strictures of the rectum as a cause of megacolon are rare they are readily detected by digital examination and easily corrected by proper dilatation

The abdomen is less distended in cases of idiopathic or acquired megacolon than in cases of Hirschsprung s disease

Soiling is common in children who have psychogenic constipation A snappy redheaded three-year-old girl obviously precocious was suspected of having Hirschsprung s disease because of severe constipation. She soiled her clothing a number of times each day The redheaded mother made no effort to conceal her frustration at not being able to force her child to conform to the rules about daily bowel evacuation While the history was being obtained this cute little nonconformist was standing in the corner straining in an effort to substantiate the mother s statements about frequent soiling She didn t have Hirschsprung s disease

Careful barium studies are necessary in both instances for a definitive diagnosis Fluoroscopic demonstration of a distal spastic narrow segment, a few inches to a foot or more in length merging into a greatly dilated colon leaves no doubt of the diagnosis Dilatation of the entire bowel all the way to the sphincter does occur in an aganglionic rectosigmoid and rectum If the history of constipation dates back to infancy it may be necessary to do a biopsy of the rectal ampulla. We prefer to do the biopsy through a transverse skin incision posterior to the rectum A generous section of circular and longitudinal muscle is removed in one piece The mucosa of the bowel is not opened. Absence of ganglion cells is proof positive of Hirschsprung s disease even though the bowel is dilated to the external sphincter

Treatment

Conservative measures in the treatment of infantile Hirschsprung s disease fail. Even though daily enemas are given the infant remains

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All this fancy talk has to be backed up by a few specific measures. Mineral oil by mouth in quantities sufficient to prevent solid impactions is number one. The amount—anywhere from 2 teaspoonfuls to 2 tablespoonfuls a day—will have to be adjusted so that the stool remains pasty. Too much oil will run through and cause soiling. Impaction must be avoided. Next it is important to try to establish a bowel evacuation pattern. Since most people believe that the proper time for defecation is once daily after breakfast, that is the time of day when the child is gently urged to go. Earnest, not maudlin, praise is given for success. Punishment for failure is disastrous. Occasionally a suppository—adult size is preferable—is necessary to stimulate action. If unsuccessful, a small enema is given. The mother has been forewarned to avoid panic and drastic measures on those days when simple measures fail and to do nothing further until the following morning. No comments or suggestions are made and no questions are asked. In the course of time—months, not weeks—the child finally catches on, moves his bowels normally and dispels gloom in the household. It is amazing how frequently older children while on a vacation or visit away from overanxious parents move their bowels daily without urging.

The *surgical* treatment of children with typical Hirschsprung's disease has become rather well standardized.

A number of days before operation is scheduled the child is admitted to the hospital for thorough bowel cleansing. It is useless to write an order for nurses to give enemas. According to the rules of the hospital, a nurse is not allowed to insert the enema tip beyond the sphincter. Consequently their enemas are completely ineffective. The surgical residents have become resigned to the unpleasant task of clearing the colon. A French catheter, no. 24 to 28, well lubricated, is carefully inserted well into the rectosigmoid. If the catheter will not easily slide into the bowel, water is injected while the catheter is being advanced. (Thus far no bowel has been punctured.) It is essential to get the catheter into the dilated portion of bowel to accomplish satisfactory lavage. Large amounts of water are not injected with the expectation that the child will expel the water and feces. The lavaging process consists rather in injecting water to and fro with a large Asepto syringe. The process of cleaning out the colon is a laborious task relished by neither resident nor patient but absolutely essential. Operation is not performed until every hard mass of fecal material has been removed. After the colon has been cleaned, neomycin is given by mouth for twenty four to thirty six hours before operation to sterilize the bowel.

The operation is performed under open mask ether anesthesia. A catheter is inserted into the bladder and a long left paramedian incision is made. The descending colon and rectosigmoid are inspected and a biopsy, without opening the mucosa, is taken from the spastic segment

somewhat distended, eats poorly, fails to gain weight and eventually becomes a poor surgical risk After a number of unsuccessful trials with conservative management of infants with aganglionosis it is now our routine to perform a colostomy.

The bowel is cleaned and sterilized with neomycin Under ether anesthesia a left paramedian incision is made The first biopsy is taken from the narrow segment of colon to prove the diagnosis, the second is taken from the most distal portion of normal-appearing colon Biopsy is performed by making a longitudinal incision in the colon A generous segment of the two muscle layers is excised in one piece without opening the mucosa—usually If ganglion cells are present in the bowel at the site of the second biopsy, a colostomy is performed at that point The loop of colon is drawn out through a separate small muscle-splitting incision Even though the loop of bowel is drawn through a small opening in the abdominal wall, it is necessary to suture the peritoneum to the bowel wall to avoid later protrusion of a loop of small bowel alongside the colon I know from sad experience that this suture is mandatory

If no ganglion cells are found in the second biopsy, a colostomy is performed in the transverse colon just proximal to the splenic flexure

The colon is opened in twenty-four hours or sooner if the infant is distended In the course of a few days the colon is transected At the same time a narrow strip of tissue is cut from the proximal end of the colostomy for further microscopic studies To distinguish between normal and aganglionic bowel requires careful study of many microscopic sections

After colostomy the child immediately improves and thrives By about age six to eight months the definitive pull-through operation is performed

Treatment of the psychically maladjusted child who has an acquired megacolon due to prolonged constipation is usually handled by the family physician or pediatrician More difficult problem children are shunted to a psychiatrist By force of circumstances the surgeon treats a few, he at least tries his simple methods of readjustment

The most difficult barrier to break through is that of convincing parents that no basic pathologic state exists (I sometimes wonder whether the huge colon which stubbornly refuses to function and acts as a reservoir rather than as a propelling mechanism has some unrecognized basic defect in nerve or muscle structure) It is emphasized that there is no fixed pattern for frequency of bowel evacuation in presumably normal people, variations from three times a day to three times a week are common Reassurance that the child is normal and will grow up and eventually overcome all difficulties is repeated again and again both to parents and child Some children—seemingly those of higher than average intelligence—glare defiance at parents and doctors as they listen to the honeyed words of reassurance

three-year-old child in serious condition was admitted to the hospital because of advanced intestinal obstruction An emergency cecostomy was done but the child died a few hours later Our pathologist Joseph Boggs was able to find no ganglion cell from pylorus to rectum Yet the child had lived three years

Most surgeons utilize the Swenson procedure as outlined above State advises wide resection of the dilated bowel but leaves at least 8 cm of the distal rectum and performs an intra abdominal anastomosis Good results are obtained with both procedures Much remains to be learned about the problem of aganglionosis of the bowel

Another biopsy is taken, not from the dilated and hypertrophied bowel immediately above the narrow segment, but from the bowel which appears and feels normal

While these sections are being cut and studied by the pathologist the operation proceeds, but no vessels are cut If the pathologist finds no ganglion cells in the second biopsy, another is taken 6 to 10 inches further up During recent years it has rarely been necessary to take a third biopsy because we have learned to recognize normal bowel Thus far in older children the clinical diagnosis of Hirschsprung's disease has always been confirmed by the absence of ganglion cells in the spastic segment

A typical pull-through operation as advised by Swenson is performed Two factors are of great importance Blood supply to the portion of colon which is to be pulled through the rectum must be adequate, the bowel must be so thoroughly mobilized that there is absolutely no tension on the suture line of the anastomosis These two factors work against each other, if the bowel is adequately mobilized, the blood supply may be insufficient, and vice versa If ganglion cells are present in the descending colon, mobilization of the splenic flexure allows plenty of bowel with an adequate blood supply If the splenic flexure is aganglionic and has to be removed, there will be some difficulty in moving the transverse colon to the rectum In five instances we have had to tie and cut the mid-colic artery, free the hepatic flexure and bring it down to the rectum Interestingly enough, the children who required such extensive resection of the colon did well after operation

RESULTS. We have performed sixty-six pull-through operations, and fifteen intra-abdominal resections as advised by David State for aganglionosis of the bowel Three deaths followed operation, all were due to peritonitis following dehiscence of the anastomosis Besides these fatal cases, a number of minor and major nonfatal complications followed leaks at the anastomosis The operation, not a simple one, nor without danger, is the best available, however, in combating this disease Those children who survived have generally had good results Problems of postoperative enteritis and incontinence have arisen from time to time, but in only a few cases Two children have not been benefited by operation and have distressing recurrence of symptoms Their abdomens are always distended, their bowel movements always liquid

A few questions remain unanswered Why does one child have serious intestinal obstruction at birth and another have only troublesome constipation for years, even though the pathology is similar in both? Do ganglion cells in the distended bowel get sick and die? Why, after identical operations, do some children have normal evacuations and a few have recurring bouts of diarrhea or constipation? How can a child survive who has no ganglion cells in the entire gastrointestinal tract? A

three-year-old child in serious condition was admitted to the hospital because of advanced intestinal obstruction An emergency cecostomy was done but the child died a few hours later Our pathologist, Joseph Boggs was able to find no ganglion cell from pylorus to rectum Yet the child had lived three years

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Congenital Atresia of the Rectum and Associated Defects

No congenital anomaly requires more thoughtful surgical care than imperforate anus or atresia of the rectum. It is so simple merely to thrust a knife into an atretic rectum and allow meconium to discharge somewhere between the buttocks and yet this thrust of the knife in the majority of cases is the beginning of an irremediable stricture eventually demanding permanent colostomy In practically every case the future of a child so unfortunate as to be born with an imperforate anus is determined by the adequacy and skill of the first operation

Diagnosis

Imperforate anus due to a membranous film at the sphincter level is a relatively innocuous but rare deformity, easily corrected by puncture and a few subsequent dilatations The only child with membranous occlusion of the rectum we have seen was cured by a student nurse The infant was admitted at age twenty-four hours with a diagnosis of imperforate anus Upon admission, according to hospital routine, a nurse shoved a thermometer into the baby's rectum The baby promptly passed a large amount of meconium, plus the thermometer, and was cured

Atresia of the rectum about 1 inch above the external sphincter is also rare, but is apt to be misdiagnosed The infant is admitted to the hospital with a clinical picture of intestinal obstruction, and, because the rectum externally appears normal and easily admits a little finger, a diagnosis of atresia of the bowel is made At operation high atresia of the rectum is found and dealt with as local conditions, such as level of obstruction and length of atretic bowel, demand

Congenital stenosis of the rectum is relatively rare and usually not discovered until the mother complains that the child is constipated and strains excessively at stool The stenosis, low in the rectum, is easily recognized and is corrected by a series of dilatations Forceful dilatation is dangerous and far less effective than persistent, gentle stretching with the best rectal dilator available—a finger

An imperforate anus is easily and promptly diagnosed If the obstetrician or even the pediatrician should forget to examine the rectum of a newborn child, he will feel a bit apologetic, but no damage will have been done when, in the first twenty-four hours after birth, the diagnosis manifests itself However, the diagnosis of imperforate anus associated with rectovaginal fistula may be missed because the infant passes meconium promptly after birth It is not uncommon to see a child with a rectovaginal fistula unsuspected until at the age of about two years it becomes impossible to toilet-train the child Rectovaginal fistulas usually appear at the fourchette, but may occur anywhere on the posterior wall of the vagina up to the posterior cul-de-sac To date we have not seen in a female infant an imperforate anus with a rectovesical fistula Rectoperineal fistulas are recognized at a glance in either sex In the male child the fistula may be very small and may open anywhere along the midline of the scrotum

When a male infant is admitted with imperforate anus, we routinely gather the available house staff to see the child After examination this question is put to them, "What is the most important point to determine next? You see that the child has an imperforate anus What else do you want to know?" Those who have not previously seen a similar case invariably want an x-ray picture to determine the position of the rectum,

or they suggest a physical examination for other abnormalities Both answers are correct. However more important at the moment is this Does the child have a hidden communication between the bladder or urethra and the rectum? To the next question "How are you going to determine whether there is a communication between the bowel and urinary tract?" the suggestion is apt to be made that the child be catheterized and that contrast medium be injected into the bladder to demonstrate the communications The diagnosis of a fistula can be simply and definitely made by examining the urine A look at the diaper flecked with meconium may give the answer A receptacle is taped to the perineum, and a specimen of urine is collected Usually particles of meconium can be seen grossly but if there is any question of identification the specimen is centrifuged and the sediment examined microscopically

Differentiation of a rectourethral fistula from a rectovesical fistula does require the use of a catheter If meconium has been identified in the urine, a very small catheter may be passed into the bladder If perfectly clear urine is obtained from the bladder or if the catheter goes directly into the bowel the fistula is in the urethra

Imperforate anus in either a male or female infant without fistula is so rare in our experience that we assume a fistula is present until proved otherwise In a children's hospital such as ours completely separated geographically from general hospitals, the most common anomaly of the rectum seen is that in which the rectal pouch ends blindly above or outside the sphincter and communicates with bladder vagina, urethra or perineum It is an assumption that relatively simple rectal deformities are treated in the hospital where the baby was born and that more complicated problems are transferred

Physical and Roentgenologic Examinations

Needless to say a general physical examination is done with special attention to the presence of other congenital abnormalities, since congenital anomalies are often multiple Congenital heart disease atresia of the esophagus and abnormalities of the genitourinary tract are the most commonly associated malformations It is important that every child with any sort of anomaly of the rectum have an intravenous pyelogram Because of distention, the pyelogram may have to be delayed until the child has well recovered after operation A pyelogram, normal or abnormal, is an important part of the record.

During the first twenty four hours after birth there is little distention of the abdomen After forty-eight hours the abdomen is usually tympanitic, and the infant has begun to vomit Since an imperforate anus is usually seen immediately after birth these infants are referred to surgery within a few hours after delivery with the anticipation of immediate

operative correction Although operation is emergent, twenty-four hours are often well spent in gathering as much information about the child as possible

Roentgenograms are routinely made in the inverted position on all infants without external fistulas A lead marker is placed over the site where the rectum should be, the child is inverted, and anteroposterior and lateral views are taken During the first twenty-four hours after birth it is not unusual to find that air has not yet found its way to the end of the colon By the second day of life the position of the end of the blind pouch is demonstrable

Operative Procedures

Before reaching for the scalpel the surgeon should have an exact mental picture of the lesion and a well formulated plan for its correction It is of the utmost importance adequately to free the rectum from surrounding structures so that it may be placed in its proper position without the slightest tension on the suture line, and to recognize and correct associated fistulas Most late complications arise from inattention to these two essential details

Surgical procedures may be divided into three more or less standard operations

1 *For a low imperforate anus ½ inch or less above the external sphincter* in an infant a perineal approach is usually satisfactory Before the infant is anesthetized the skin over the region where the rectum should be is snapped with a finger or touched with a pin Reflex contraction of the sphincter muscle will demonstrate its location, which is marked with ineradicable cross scratches with a pin Demonstration of the position of the spincter is necessary before all operations for imperforate anus

After the child has been anesthetized and placed in the lithotomy position a catheter is inserted into the bladder It sometimes requires much patient persistence to insert a catheter, but by no means should the operation begin until a catheter is in the bladder One never knows how near the atretic rectum the urethra may lie, in fact, the two structures often are in close juxtaposition It is reassuring during the operation to be able to feel the catheter and thus avoid injury to the urethra A longitudinal incision is made through the skin over the previously demonstrated sphincter, and the fibers of the muscle are gently spread If the end of the bowel is low, it may be grasped, freed from surrounding structures, pulled down through the sphincter, opened and sutured to the skin edges *There may be no tension on the suture line* If the bowel is not against the skin over the rectum, it may be necessary to make a

longer skin incision and split the sphincter to allow room for adequate dissection

Male infants having a perineal fistula and a low rectal pouch are treated in the same manner as those having a low rectal pouch without a fistula The fistula is dissected out, and the anterior surface of the rectum is freed the posterior wall is easily freed with little dissection The fistula is amputated, and the end of the bowel is sewn to the skin with silk sutures In another group of cases there is a small opening immediately anterior to the normal position of the rectum Dilatation of such openings will be unsatisfactory An incision is made posteriorly the muscle and a constricting band are cut and the rectal mucosa is well separated laterally and somewhat proximally The mucosa is then drawn down and its edges are sewn transversely to the skin

2. *An imperforate anus with a rectovesical fistula* is repaired by a combined abdominoperineal approach

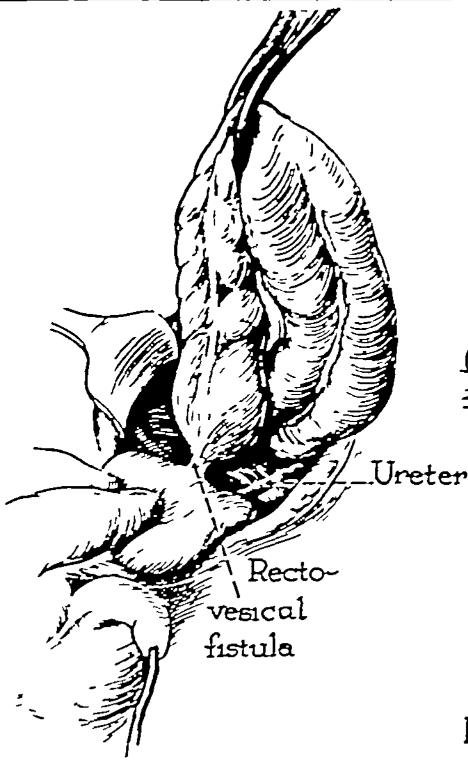
The infant is placed in the lithotomy position with his buttocks extending slightly over the end of the table The bent knees are supported by small sandbags on each side of the child s hips The entire abdomen perineal region and the inner surface of the thighs are surgically prepared A catheter is inserted into the bladder and fixed so that it cannot slip out during operation A small longitudinal incision is made through the skin over the predetermined position of the sphincter The sphincter muscle and the loose tissue in the lower pelvis are gently spread with a hemostat.

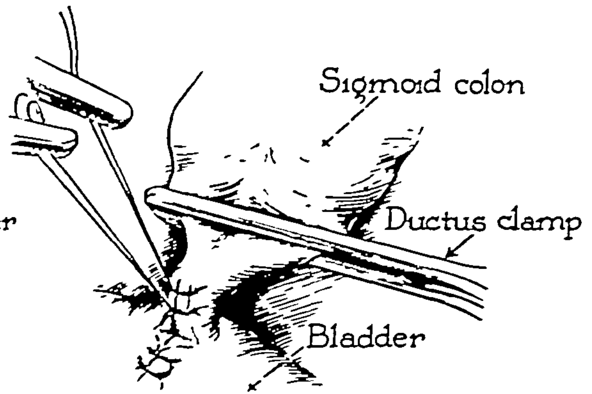
A left suprapubic paramedian incision is made The rectosigmoid is identified, freed from its peritoneal and pelvic attachments and if greatly distended with air and meconium is deflated. To prevent refilling of the rectosigmoid, a toothed coarctation clamp is applied high on the sigmoid The rectal pouch is dissected free from its lateral pelvic attachments and from the hollow of the sacrum.

The bladder is retracted forward, the rectum backward to identify the connection between the two The fistula, usually found at the base of the bladder in the region of the trigone, is divided bit by bit, and each advancing cut is closed with interrupted sutures of surgical gut on the bladder side and silk on the bowel side The silk sutures are cut long and used for gentle traction

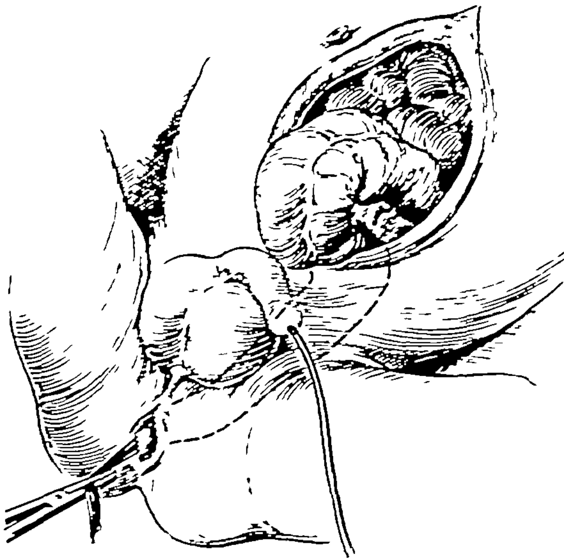
A curved hemostat is now inserted through the previously made incision in the perineum. The sutures on the end of the bowel are caught and the bowel is pulled down the proper distance so that its end can be sutured to the skin without tension In all instances the opening is made large enough to admit the operator s little finger The abdomen is closed in layers and the infant returned to an oxygenated crib The catheter is left in the bladder for five to seven days

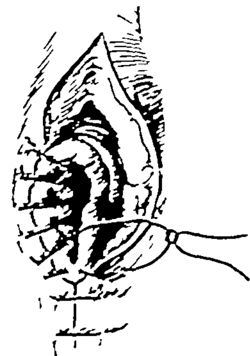
Rectourethral fistulas usually lie just distal to the prostate and are





Detail of division and suturing of fistula and bladder





Recto-sigmoid partially sutured to skin

in a sort of no-man's land Although the fistula itself may be approached more easily from below it is difficult to free the rectum sufficiently for proper positioning and suturing through a perineal incision The combined abdominoperineal approach is usually used Completely satisfactory closure of the urethral side of the fistula from below or above is difficult and seems not too important so long as the urethra lies against normal bowel wall and a catheter is kept in the bladder for at least a week

3 *Repair of imperforate anus complicated with rectovaginal fistula* When originally describing the technique of repair of this deformity we advised operation during the neonatal period That was a mistake The operation should be delayed until the child is about six months of age by this time the hypertrophy of the bowel wall which will have occurred will facilitate dissection in the proper line of cleavage During these months the bowels move through the vaginal fistula without difficulty if the diet is adjusted for free evacuation Dilatation of the fistula is occasionally necessary To avoid impactions we instruct the mother that under no circumstances shall she allow the baby to go more than twenty four hours without adequate bowel evacuation An enema can easily be given with a catheter inserted through the fistula.

During the forty-eight hours preceding operation the bowel is sterilized with adequate doses of neomycin given by mouth. The evening before and the morning of operation the bowel is irrigated with sterile salt solution until clear Woe unto the resident who doesn't have that bowel absolutely clean and free from irrigating fluid!

The technique of operation is shown in the illustration on page 208

An important step in this operation consists in placing the bowel *behind* the *intact* median raphe

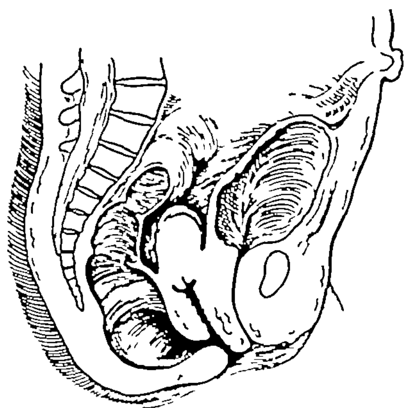
In case the rectovaginal fistula is situated high in the posterior wall of the vagina, an attempt is made to free the rectum through a perineal incision but provisions are made for an abdominoperineal approach should that be necessary

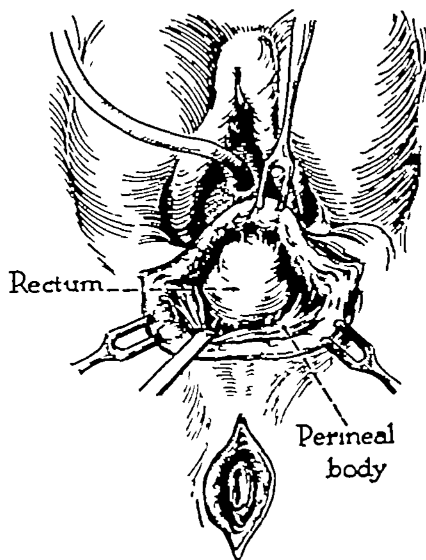
A rectoperineal fistula in the female infant is handled in the same way except that the curved transverse skin incision is made around the fistula and to each side as far as necessary for adequate exposure

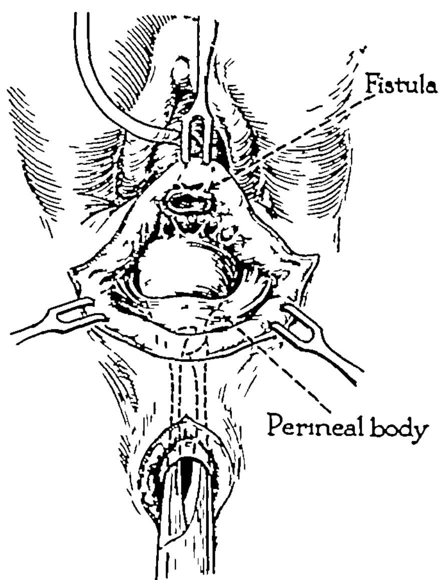
The catheter is removed after the operation has been completed.

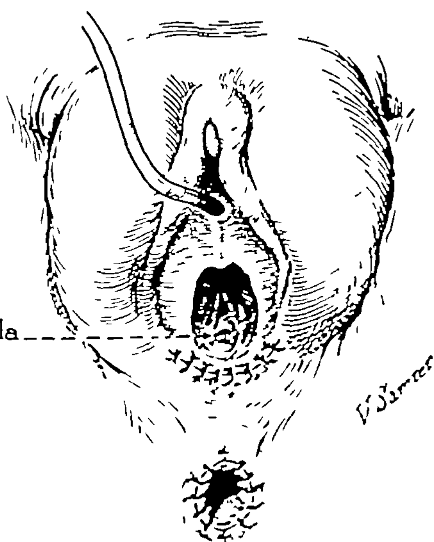
Postoperative Care

Immediate postoperative care is the same after all operations for imperforate anus After the child has fully regained consciousness he is placed on his back, and both legs are suspended as in Buck's extension for fractured femurs except that the suspension is more simply effected Adhesive strips, applied to both sides of both legs are attached with


Diagram of fistula


Rectum
Perineal body


Fistula
Perineal body
Allis forceps through sphincter


Closed fistula

pieces of rubber tubing to a rope tied across the top of the crib The object of suspending the legs is to allow urine and feces to escape onto a diaper placed beneath the buttocks rather than into an occlusive soggy diaper Better healing occurs when the wounds are exposed to the air

During the immediate postoperative period the condition of the suture line is observed daily If it gives way the reason is almost invariably the fact that the bowel was not sufficiently freed and that there was consequent tension on the suture line Immediate reoperation is necessary If not corrected the raw area between the end of the bowel and the skin will be the site of an iron hard fibrous stricture which will not yield to gentle or even forceful dilatation

One teaspoonful of mineral oil is given each night for about one week to avoid the possibility of fecal impaction As soon as healing has occurred the oil is stopped and is not resumed at any time thereafter Oil tends to run through the bowel in these patients and causes continuous soiling and excoriation of the buttocks Dilatation of the rectum will always be necessary but is delayed until about the tenth day to allow firm healing of the suture line and is then most satisfactorily done with the tip of a well lubricated centrifuge tube

The child is discharged from the hospital about two weeks after operation Continued and determined follow up attention for years is of the utmost importance For those patients who live beyond the suburbs the help of the pediatrician or family physician is essential

The mother is instructed to dilate the infant s rectum every other day with her little finger sheathed in a finger cot. When she is able to insert her index finger beyond the first joint, dilatations are discontinued

The diet must be so regulated that the stools are pasty Too loose stools result in constant small evacuations and raw buttocks Constipation during infancy is equally bad because impactions are difficult to deal with. The mother is told that if the baby does not pass some formed stool every day an enema must be given

For about two years as the infant thrives the mother is happy and unconcerned about the fact that the baby s bowels move somewhat irregularly As the time for toilet training arrives, conferences with the mother are in order Female infants who have had repair of rectoperineal or rectovaginal fistulas and the infants who have had correction of a simple imperforate anus can be toilet-trained usually between their second and third years Eventually most of these children have normal control of bowel evacuation. Male infants who have had an abdominoperineal repair cannot be completely toilet trained—let s face it—until they are about five or six years old and themselves begin to take interest in keeping their pants clean. Parents are terribly disturbed about the fact that their boy still soils his pants It is explained to them that the lower segment of the bowel with its important nerve supply is missing and that

consequently the reflex arc which signals need for bowel evacuation is interrupted These boys are put on such constipating diets that their stools are constantly firm They may have accidents, but not offensive ones, from time to time just a little hard ball rolls out of their pants leg The passage of firm constipated stools subconsciously helps to educate the evacuating mechanism The importance of a daily bowel movement is emphasized

When the boy goes to kindergarten, the mother is instructed to give him a small enema if his bowels do not move spontaneously, to avoid accidents at school and the psychologic trauma of being ridiculed by his playmates Slowly the child begins to use his external sphincter, levator muscles and buttocks for the control of evacuation A definite time for stool is established, and the boy is coached to grunt or bear down When successful, he avoids the enema and receives praise, when unsuccessful, he gets an enema or a suppository, but is never criticized By the time the boy is five to six years old he usually has developed sufficient control to move his bowels once daily The only time he will have trouble is during attacks of diarrhea Eventually these boys get along fairly well, although not one of them has a perfect rectum that functions like an original model

The cause of most difficulties in later years, even after suitable reconstruction of the rectum in infancy, is lack of observation and instruction to parents Because the child is kept on a constipating diet, impactions are apt to occur Parents report that their boy was getting along fairly well, but missed moving his bowels for a few days and began soiling his pants many times each day Because the bowels are moving the parents don't suspect impaction, and I regret to add that the physician sometimes fails to confirm it by inserting a finger into the rectum Removal of impactions often extending far into the colon in neglected cases is accomplished with soap suds enemas given twice daily until every mass has been discharged Again instructions are repeated

Colostomy

Temporary colostomy is performed only on those infants who are in poor condition because of prematurity or multiple anomalies, permanent colostomy is unavoidable for those who have such extensive deformities of the buttocks that construction of a rectum is impossible For those patients upon whom a later construction of a rectum is planned, it is important to place the colostomy in the left portion of the transverse colon so that at the second operation the distal segment can be brought down without the mechanical handicap of fixed bowel

The usual technique of colostomy is used, but it is important to divide the colon completely in order to minimize flow of feces into the

distal segment If conditions demand a permanent colostomy it is well to remove the blind end of the colon distal to the colostomy to avoid accumulation of fecal masses

After previous colostomy plastic procedures on the imperforate anus should be done before the infant is nine months old so that all post operative dilatations can be completed while the infant is easily man ageable and before the age of known memory of pain Unfortunately somewhere along the line colostomies have been advised for most in fants with imperforate anus and plastic work on the rectum when the child is two to five years old Those who have had to do postoperative rectal dilatations on children of this age will not soon forget the ordeal the infant screams and fights the mother cries and the surgeon feels like a cruel monster Get the unpleasant task completed before the infant remembers from one visit to another the unpleasantness of rectal dilata tion Why not make the opening large enough so that dilatations will not be necessary?—For the simple reason that in so doing the objective of a normal or near normal rectum is defeated

This conclusion is inescapable In general atresia of the rectum is more poorly handled than any other congenital anomaly of the newborn A properly functioning rectum is an unappreciated gift of greatest price The child who is so unfortunate as to be born with an imperforate anus may be saved a lifetime of misery and social seclusion by the surgeon who with skill diligence and judgment performs the first opera tion on the malformed rectum

Rectal Bleeding

Parents know that bleeding from the bowel is a sign of cancer and it is natural that when a child passes blood, they apprehensively seek medical aid. One may promptly allay their anxiety. During the seventy-five-year existence of the Children's Memorial Hospital no case of primary adult type of cancer of the bowel has been seen in children below sixteen years of age. Cases have been reported in the medical literature but are extremely rare. When a mother reports that her infant or child has passed blood, the common causes of such bleeding are considered.

Etiology

Fissures in the rectum are seen in infants below one year of age—rarely in older children. A few drops of bright red blood appear on the outside of the stool. The infant is apt to be constipated and cries during defecation. Upon rectal examination one easily detects the tiny fissure

in the mucosa A bit of mineral oil by mouth to soften the stool and some moral support are all that is needed in the way of treatment

Hemorrhoids—real, ugly, bulging, blue varicosities as one sees in an adult—do not occur in children I have seen two children with hemorrhoids, in one instance they were part of a hemangioma, in the other they were a result of portal hypertension

Rectal polyps—usually single, occasionally multiple—are relatively common in children from about two to six years of age A little bright red blood is seen on the outside of the stool The mother reports that sometimes after defecation the child has to go again and this time passes a small blood clot, and when she adds that she has seen what looks like a cherry come out of the rectum, the diagnosis is made Fortunately, polyps in children are usually confined to the rectum and are easily felt Surgical removal with a snare or cautery is effective Recurrences are rare

When the history is typical of polyps, but none can be felt on digital examination of a thoroughly cleared rectum, proctoscopic examination is indicated An enema is given the night before, and another one two hours before examination It is preferable in a cooperative child supported by a fairly liberal dose of sedative to do the proctoscopic examination without general anesthesia The conscious child, by straining a bit (consciously or unconsciously), may push down a pedunculated polyp which otherwise might not be seen One should be prepared to grasp such a high-lying polyp quickly with an instrument before it moves out of the field

Polyps above the rectosigmoid may or may not be associated with the passage of bright red blood Fluoroscopic and roentgenologic localization of the polyp high in the rectosigmoid may be difficult If bleeding has been persistent, diagnostic pursuit must be equally persistent A polyp that cannot be reached by proctoscope will have to be removed transabdominally Only if the bowel has been thoroughly cleared will it be possible to find the polyp Occasionally a child will have unexplained secondary anemia due to a colonic polyp unsuspected because the child has passed no visible blood Roentgenographic demonstration of such a polyp is an ego-inflating success A child who has persistent secondary anemia without demonstrable cause, and whose stool contains occult blood while he is on a strictly meat-free diet, is entitled to surgical exploration even though repeated roentgenographic and proctologic studies of the gastrointestinal tract are negative

Melanin spots in the vermilion border of the lips in a child who has rectal bleeding are practically diagnostic of polyposis of the bowel (Peutz-Jegher syndrome)

Multiple polyposis is a quite different story A history of familial tendency to the disease is common The child is apt to pass blood with every stool and is often anemic Numerous polyps are felt in the rectum,

and double contrast films will demonstrate many—often hundreds—in the colon

What to do surgically for a six to eight year-old child with multiple polyposis is difficult to decide It is especially desirable in a child of this age to preserve the sphincter and normal bowel evacuation at the same time it is essential that all polyps and this means most of the colon be removed

If it is decided to preserve normal bowel evacuation the rectum is freed of all polyps by fulguration With the aid of a proctoscope and under general anesthesia at a number of sessions all the polyps for a distance of at least 20 cm are removed After the rectum has healed and has been checked to be sure no polyp remains colectomy is performed and the ileum is anastomosed to the upper end of the rectum The child will have considerable trouble after this operation with loose stools for a few months but eventually as the distal ileum takes over the function of the colon he will develop fairly normal bowel habits

The importance of having the child returned for frequent proctoscopic examinations for recurring polyps in the remaining rectum cannot be overemphasized. Occasional polyps are apt to appear and will have to be removed as they arise

At what point if polyps continue to recur does one give up and advise removal of the distal portion of the rectum? So long as one can keep the rectum free of recurrences it seems justified to give the child a chance for normal living. At the same time, if recurrences are frequent, it is foolhardy to fight a losing battle and risk the chance of carcinomatous degeneration

One of two surgical courses is open when all are agreed that the rectum must be removed The remaining segment of rectum is resected including all the mucosa to the rectoperineal line The ileum is then drawn through the sphincter where it is allowed to seal itself to the raw surfaces The excess is trimmed off later This operation has been tried only once, and oddly enough the child now has fairly good bowel control. In general, however the alternative procedure of completely resecting the rectum and doing an abdominal ileostomy is most frequently chosen Certainly an abdominal ileostomy properly fitted with a Rutzen bag is less objectionable than an incontinent ileostomy between the buttocks It can be stated unequivocably that in the treatment of multiple polyposis there may be no compromise All polyps must be removed, or eventually carcinoma will develop

Chronic ulcerative colitis is a dread disease The etiology is still completely obscure Infection in the bowel is almost certainly secondary to the basic disease Emotional maladjustments have been suggested as causative but it appears that psychogenic factors aggravate rather than initiate the process

Ulcerative colitis typified by the usual frequent evacuation of mucus, blood and pus may appear in infants from three to six months of age, but the symptoms and findings are less severe than in older children. At proctoscopic examination, gently done under general anesthesia, one sees on the mucosa a grayish, mucoid, adherent layer which, when removed with a swab, leaves a congested reddened mucosa dotted with punctate hemorrhage. Ulcers are seen in older children and in more advanced cases.

A barium enema in an infant will show relatively little narrowing of the lumen of the colon, in contrast to the picture in the older child, whose colon has lost all haustrations and looks like a curved piece of garden hose.

The treatment of ulcerative colitis varies according to the past experience of the physician and the attitude of the medical staff. A few children, all will agree, will require ileostomy as a lifesaving procedure. Except for the chronically severe and advanced cases, obviously incurable medically, all possible means of conservative management are in order. Diet, vitamins, drugs, rest, cortisone, antibiotics, time, optimism, loving care—all are conscientiously applied to this disease in the hope of averting an ileostomy. If an ileostomy becomes necessary, it should be done as simply as possible. The proximal end of the ileum is brought out through a stab wound in the right lower quadrant and so placed that a Rutzen bag can later be adjusted without difficulty. The distal end of the ileum is brought out through the exploratory incision or through another small wound well away from the ileostomy stoma so that it will not interfere with the wearing of a bag. One ileostomy and three permanent colostomies have been placed in the mid-abdomen at the site of an excised umbilicus. Insufficient experience disallows conclusions, but it does appear in each case that the Rutzen bag fits better in this position and is less apt to loosen during exercise.

Accustoming the child to the ileostomy bag is a task of patience and forbearance. Diet will have to be controlled, and the skin about the ileostomy will have to be protected from erosion by intestinal juices. Aluminum hydroxide paste in the early days after ileostomy is moderately effective. Later, nothing is more effective than an ileostomy bag so fitted that the edges of the rubber flanges cover the skin to the very border of the ileostomy. A neglected ileostomy is a torturous thing. A six-year-old boy with an ileostomy—the skin about it raw, red and oozing—was admitted to the hospital as a nervous and physical wreck. By placing him in the prone position on a Bradford frame so that all discharges from the intestine flowed directly into a pan, healing of the skin was finally accomplished.

One doesn't take from parents hope that eventually continuity of the bowel may be restored. No directions can be given for a suitable time

for this procedure Certainly all discharge of mucus pus and blood from the rectum must have been absent for years and the mucosa of the recto-sigmoid must have regained normal appearance before closure is contemplated Parents who urge closure of the ileostomy are warned that the procedure may be followed by recurrence of symptoms Some children will be doomed to a lifetime ileostomy At times the colon is so badly diseased that total colectomy is necessary

Before leaving this subject it seems appropriate to make reference to *idiopathic bleeding* from the rectum in early infancy An infant who does not present the usual picture of infectious or specific diarrhea may pass large quantities of diluted blood and mucus The pediatrician is concerned about the amount of blood passed thinks some specific bowel lesion accounts for it and calls in a surgeon The infant does not look particularly ill or dehydrated as is the case in severe diarrhea of infancy yet the amount of red staining of the diaper is disturbing Finally the surgeon capitulates and operates—to his sorrow We have seen approximately twenty such cases during the past ten years and occasionally have operated and been humbled by finding no pathologic condition

Such nonproductive exploration makes the surgeon eligible for membership in the S O B Club The abbreviation does not, as some might suppose refer to the usual interpretation it stands for Surgeon of Bleeders All the general surgeons at our hospital have finally won a membership the pediatricians involved receive associate memberships This membership is a reminder that diffuse, watery bleeding from the rectum in infants during the first few weeks of life is part of some unknown syndrome From visits to other children s hospitals it has been learned that similar societies there would also have a sizable membership Medical treatment with vitamins of all kinds lots of vitamin K, changes in formula, and above all, time eventually lead to or are associated with recovery

Inguinal Hernia

Repair of an inguinal hernia is the most common operation in the field of general pediatric surgery For sixty years English surgeons supported by their Australian confreres, have advocated simple ligation of the sac as the treatment of choice Nevertheless much difference of opinion still exists and the following statements are often heard Truss support is sufficient, operation should be delayed until the child is two to four years old inguinal hernias disappear spontaneously complicated methods of repair are essential. Nonsense! The cause of inguinal hernia is persistence or inadequate closure of the processus vaginalis peritonaei. The logical treatment, therefore is simple removal of that offending sac.

If these statements sound dictatorial, listen to what Ferguson said with an oratorical flourish in 1899 "Tearing the cord out of its bed is without anatomic reason to recommend it a physiologic act to suggest it nor [does it give] brilliant surgical results to justify its continuance. Leave the cord alone, for it is the sacred pathway along which travel elements indispensible to the perpetuity of our race"

In a full term infant the processus peritonaei normally is closed Parents often believe that excessive crying is the cause of their child's hernia If the processus peritonaei or sac is completely obliterated, crying will not cause a hernia, if, however, the smooth surfaces of the sac are merely adherent, but not fibrotic, crying or straining may suddenly separate them and force the bowel into the inguinal canal or all the way into the scrotum, depending upon the extent to which the sac has been opened Books and articles ascribe all sorts of names to variations of inguinal hernia Actually, inguinal hernias in children are that and nothing more—they just vary in size Hernias in infants and in children through the early teens are with few exceptions indirect Direct and femoral hernias are so rare that they will not be included in the discussion

Diagnosis

A diagnosis of inguinal hernia is usually made by the parents Confirmation is made by the physician who finds in the inguinal canal a bulge which can easily be reduced At times it is difficult, if not impossible, to be sure that a hernia exists The mother gives an uncertain history of having seen a lump in the groin once or twice Even though the child cries during the examination, no hernia appears Absolutely no information will be gained from trying to slide a finger in the ring as is done in adults Palpation of the inguinal area with a sideways rubbing motion of the first finger parallel with the inguinal ligament will impart a feeling of rubbing silky surfaces together if a sac is present This maneuver requires some practice Whether or not to advise bilateral herniotomy when an obvious hernia is present on only one side is still a matter of dispute If a hernia is present on the left side, special attention is given to examination of the opposite side because 80 per cent of hernias occur on the right Even though the examiner cannot with certainty palpate a sac on the right side, a hernia on the left justifies advising bilateral herniotomy We do not advise routine bilateral herniotomy when a demonstrable hernia is present on the right side

Female infants often have an ovary and/or tube in the inguinal canal In a recent study of 200 hernias in female infants and children we found that 21 per cent were of the sliding variety, involving various parts of the genital tract The histories of 29 per cent of the patients contained notations of various symptoms observed by the mother, such as excessive crying, colic, irritability, pain, vomiting, constipation and feeding problems The symptoms largely disappeared after herniotomy

HYDROCELE A hydrocele is a collection of fluid in a portion of unobliterated processus peritonaei It may be confined to a segment of the cord or may extend all the way to the testicle Numerous times it

has been pointed out that a hernia in children is practically always associated with a hydrocele. A small opening in the distal portion of the hernial sac allows fluid to collect in the second compartment and the condition is labelled hydrocele. If the parents report that the round swelling in the scrotum is largely gone in the morning or if pressure on the hydrocele reduces its size, a hernia, although it may not be demonstrable, is unquestionably present. Small hydroceles of the cord are relatively common during infancy and often disappear spontaneously in infants below six months of age. In female infants the corollary is a hydrocele of the canal of Nuck. It is not wise to establish a diagnosis of hydrocele by needle aspiration of fluid. One of my confreres tried and was more than chagrined to withdraw air and intestinal content. The child had an emergency operation and all went well.

INCARCERATED HERNIA Incarceration is the most feared complication of inguinal hernia. A detailed study of hernias in infants and children brought out the fact that at age 2 months 23 per cent of infants with hernias either were admitted with incarceration or had a history of previous incarceration. This high rate falls off in a sloping curve to 5 per cent at 8 months of age and continues at about that level during childhood.

Reasonable attempts should be made to reduce an incarcerated hernia in an infant before proceeding to operation because of the difficulty in dissecting out and properly closing an edematous sac which has about as much strength as wet tissue paper. With the baby held up by the legs so that only his head touches the table, firm pressure on the mass will usually effect a reduction. The crying baby may be quieted and led into a cooperative attitude by giving him a pacifier reinforced with sugar.

If reduction is not promptly accomplished by this maneuver, a sedative is given, adhesive strips are applied to the baby's legs, and enough traction is made by weights over the end of the crib to elevate the child's buttocks. Within an hour or two reduction practically always occurs either spontaneously or with the aid of slight pressure on the mass. Elective operation is performed later. Obviously, attempts at reduction must be tempered with good sense. It can happen that a resident who has just reduced a hernia will receive a dark look from the attending man who thought it should have been operated upon at once.

Treatment

When a diagnosis of inguinal hernia has been made in infant or child, operation is advised regardless of age. If the infant is otherwise normal and is gaining weight, there is no reason to delay surgical correction. Such advice is often questioned by parents because of rumors that an infant cannot tolerate anesthesia and operation, or that the hernia

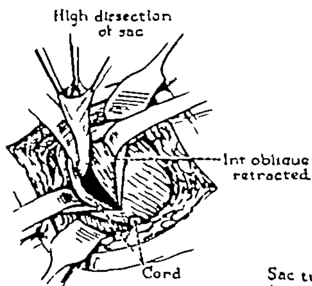
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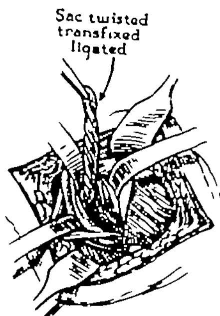
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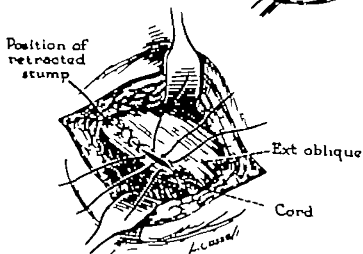
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High dissection of sac
Int oblique retracted
Cord


Sac twisted transfixed ligated

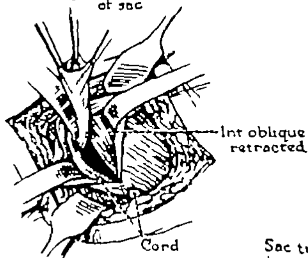

Position of retracted stump
Ext oblique
Cord

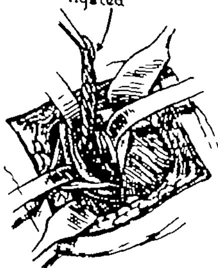
will get well spontaneously Pediatricians are becoming convinced that the worrisome problem of an inguinal hernia in an infant can be solved quickly and safely by operation

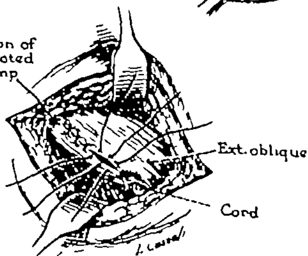
Infants who are underweight, poor feeders and sickly, or have other deformities are temporarily fitted with a truss For tiny infants no truss is altogether satisfactory Yarn trusses, popular for years, reassure the mother, but do the baby little good The least unsatisfactory truss for infants who must wear one is an elastic belt with perineal strap which holds an inflated rubber pad over the inguinal region For older children who are unfit for surgery a spring truss is as good as any, but is difficult to keep in place

The operation is performed under ether anesthesia A transverse incision is made in the inguinal region—in infants in the ever-present suprapubic crease The external oblique is split along its fibers, but the external ring is not opened Obviously, if the external ring is not opened, it doesn't have to be sewn up later, and the danger of closing it too tightly is eliminated One of the prime causes of hematomas of the scrotum after operation, I am convinced, is too snug closure of the external ring and consequent obstruction of the venous circulation The sac is grasped with a hemostat and lifted up without elevating the structures of the cord With a finger in the open sac as a guide, all the structures of the cord, including the tiniest vessels, are gently and thoroughly freed from the paper-thin sac until one sees properitoneal fat in the region of the internal ring After the sac has been completely freed its interior is inspected, and it is then twisted until the last turn is flush with the parietal peritoneum The sac is transfixed with a 000 silk suture and securely tied The excess of sac is cut away, and if the dissection has been complete, the stump will retract under the internal oblique muscle The external oblique is closed with a few interrupted sutures and the skin with silk In infants who have not been toilet-trained the skin edges are coapted with interrupted subcuticular sutures of 6-0 white silk White silk rather than black is used for the subcuticular sutures because the latter are apt to show through the skin "Doctor, what are those dirty spots under the incision?" A tiny strip of gauze is laid over the incision and covered with waterproof adhesive tape The child is discharged from the hospital the following morning No restrictions in activity are advised for children below five years of age Older children are cautioned against violent exercise for a couple of weeks

The transverse incision looks much better immediately after operation and is less apt to result in keloid formation In infants the transverse incision is especially desirable because the edges of the wound practically fall together When the mother returns with her child for check-up examination, she is apt to say, "Oh, what a beautiful scar" (Very important to the reputation of the surgeon are the complimentary remarks


High dissection of sac
Int oblique retracted
Cord


Sac twisted, transfixed ligated


Position of retracted stump
Ext. oblique
Cord

Mother makes to her friends at the Tat and Chatter Circle about the tiny hairline scar)

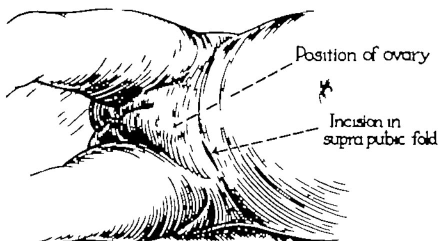
A large persistent hydrocele is approached by the same transverse inguinal incision It is opened, evacuated, dissected from the structures of the cord and cut off The distal end is left open (The so-called bottle operation is never performed in a child) The hernial sac is then sought for—it will almost invariably be found—and treated as outlined above

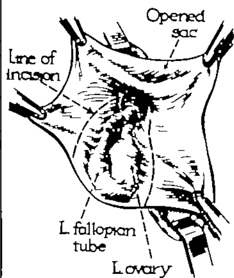
Simple inguinal hernia in a girl is repaired in the same way as in a boy, but the sliding hernia of ovary and/or tube requires a different technique After the sac has been freed from the fibers of the cremasteric muscle and connective tissue, care is exercised to avoid damage to the ovarian and uterine vessels in the posteromedial portion of the sac The sac is opened, and if the tube and/or ovary with their blood vessels form part of the sac—typical of sliding hernia—the following procedure is carried out A flap is fashioned from the side of the sac to which the adnexa and their vessels are adherent by making an incision in the sac on each side of the vessels and parallel with them This tongue or flap of sac with its attached ovary or tube and vessels is then folded through the neck of the sac into the peritoneal cavity A purse-string suture of 000 silk is so placed in the remaining portion of the sac that when it is drawn up and tied, it will completely close the sac without encroaching upon the vessels to ovary or tube The redundant portion of the sac is removed and the wound closed as above

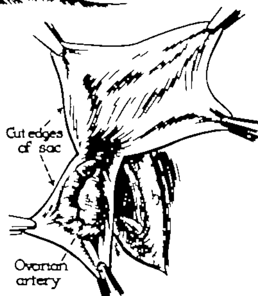
A word of advice from one who has learned the hard way It is now an ironclad rule at our hospital that whoever takes the history and makes the physical examination on a child admitted for herniotomy has one of the parents write and *sign* a statement on the clinical record as follows, "My son (daughter) has a hernia on the ——— side"

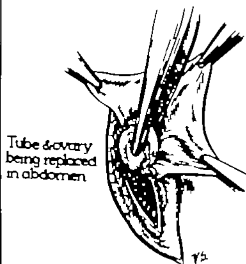
Results of surgical treatment of inguinal hernia are most satisfactory Recurrences are rare—less than 1 per cent Morbidity and mortality are low The only death in a series of approximately 2000 herniotomies occurred in a 4-pound premature infant who was admitted to the hospital *in extremis* with a strangulated hernia At operation the bowel was released and was questionably viable Because the infant was near death, replacement of the bowel in the abdomen seemed better judgment than resection At autopsy on the following day rupture of the weakened bowel and early peritonitis were found

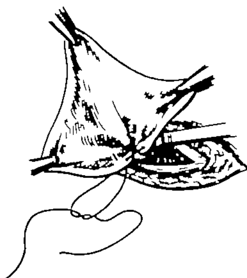
After operation parents are grateful that their worries about a troublesome hernia have been dispelled They can now go out weekends without worrying that their child left with Grandma or a nursemaid is going to have an incarceration requiring emergency operation


Position of ovary
Incision in
supra pubic fold


Opened
sac
Line of
incision
L. fallopian
tube
L. ovary


Cut edges
of sac
Ovarian
artery


Tube & ovary
being replaced
in abdomen



Mother makes to her friends at the Tat and Chatter Circle about the tiny hairline scar)

A large persistent hydrocele is approached by the same transverse inguinal incision It is opened, evacuated, dissected from the structures of the cord and cut off The distal end is left open. (The so-called bottle operation is never performed in a child) The hernial sac is then sought for—it will almost invariably be found—and treated as outlined above

Simple inguinal hernia in a girl is repaired in the same way as in a boy, but the sliding hernia of ovary and/or tube requires a different technique After the sac has been freed from the fibers of the cremasteric muscle and connective tissue, care is exercised to avoid damage to the ovarian and uterine vessels in the posteromedial portion of the sac The sac is opened, and if the tube and/or ovary with their blood vessels form part of the sac—typical of sliding hernia—the following procedure is carried out A flap is fashioned from the side of the sac to which the adnexa and their vessels are adherent by making an incision in the sac on each side of the vessels and parallel with them This tongue or flap of sac with its attached ovary or tube and vessels is then folded through the neck of the sac into the peritoneal cavity A purse-string suture of 000 silk is so placed in the remaining portion of the sac that when it is drawn up and tied, it will completely close the sac without encroaching upon the vessels to ovary or tube The redundant portion of the sac is removed and the wound closed as above

A word of advice from one who has learned the hard way It is now an ironclad rule at our hospital that whoever takes the history and makes the physical examination on a child admitted for herniotomy has one of the parents write and *sign* a statement on the clinical record as follows, "My son (daughter) has a hernia on the ——— side"

Results of surgical treatment of inguinal hernia are most satisfactory Recurrences are rare—less than 1 per cent Morbidity and mortality are low The only death in a series of approximately 2000 herniotomies occurred in a 4-pound premature infant who was admitted to the hospital *in extremis* with a strangulated hernia At operation the bowel was released and was questionably viable Because the infant was near death, replacement of the bowel in the abdomen seemed better judgment than resection At autopsy on the following day rupture of the weakened bowel and early peritonitis were found

After operation parents are grateful that their worries about a troublesome hernia have been dispelled They can now go out weekends without worrying that their child left with Grandma or a nursemaid is going to have an incarceration requiring emergency operation

Undescended Testicle

No subject is more hotly disputed today than the management of cryptorchidism At any panel discussion this simple question, "At what age do you advise surgery for unilateral undescended testes for bilateral undescended testes?" receives widely divergent answers. Two articles on cryptorchidism appeared in one issue of the Journal of the American Medical Association in 1957 One stated emphatically that the unilateral undescended testicle should be operated upon between ages five and six years and that bilateral undescended testicles should be operated upon between ages two and three years The following article (the editor Austin Smith, with subtle humor placed the articles in direct sequence) vehemently stated that practically all undescended testicles eventually and unaided find their rightful position in the scrotum and that operation (rarely necessary) should be delayed until the boy is sixteen or seventeen years old.

The reason for so much heat and so little light on this subject is the fact that few observers have studied sufficient number of cases of

of undescended testicle and write a letter to the referring physician that by good fortune it was possible to push the testicle in the scrotum and that operation may be delayed—one need not add, indefinitely

Treatment

Guided by a wide variety of suggestions obtained from medical literature and by our own personal experience with management of uni lateral and bilateral cryptorchidism, we have come to the following conclusions, subject to change without notice

Operation for unilateral cryptorchidism is delayed until the child is about nine to eleven years old, for these reasons A fair number of testes descend spontaneously during this time it has not been proved that early operation furnishes any advantage for more normal testicular development, there is less likelihood of fatally damaging the spermatic blood vessels during operation on the older child the larger testicle is more easily held in the scrotum than the pea sized testicle of a three-year-old child finally the operation by the surgeon of restricted experi ence in this field is easier in the older child and, from the standpoint of preserving the blood supply to the testicle, safer

Exceptions to any rule arise. Usually the hernia associated with an undescended testicle is small and symptomless, but should it be large and troublesome operative repair of the hernia and simultaneous placement of the testicle in the scrotum are indicated at any age. A child with an undescended testicle cannot wear a truss

Parents often ask this intelligent question, "If one testicle is sufficient for fertility why do you advise operation on the other?" The answer is, "Mainly because it is good insurance to have a second testicle in the scrotum. *Only if it is in the scrotum will it be able to form normally* active spermatozoa Besides the hernia can be repaired at the same time. For psychologic reasons also, it is well to have both testicles in their normal position."

Hormonal treatment is no longer given before ten or eleven years, and then only before operation to fat boys who have a tiny penis and pea sized testicles There is little question that gonadotropic hormones make the testes enlarge and at times promote their descent into the scrotum. It seems logical to conclude that if hormones are effective, the child s own hormones later will accomplish the same desired result. During the mid thirties members of our staff gave gonadotropic hormones to a considerable number of boys with cryptorchidism. A few children, some only two years old, consequently developed pubic hair startling enlargement of the penis and grown up ideas. After one of these boys a six year-old, had been observed making meaningful passes at a nurse it seemed time to discontinue medication capable of instilling such pre-

bilateral cryptorchidism, operated upon and not operated upon, for enough years to answer the one final and vital question, "Is the adult male fertile?" Since one testicle is sufficient for fertility, no conclusions can be reached concerning the effectiveness of unilateral operations

Examination

The ocurrence of undescended testes in boys of grade school age, reported in medical literature, varies from 2 to as high as 9 per cent The former figure is probably nearly correct, the latter is obviously too high The reason for such a discrepancy in figures can lie only in the definition of what constitutes cryptorchidism An undescended testicle is one which upon examination and re-examination under ideal conditions cannot be brought into the scrotum—not even into the upper part of the scrotum

Before asking the reluctant, sensitive boy to take down his pants, rapport must be established Examination of a wiggling youngster constantly bending over and crossing his legs is bound to be unsatisfactory The room must be warm, and, above all, the examiner's hands must be warm Fear as well as cold stimulates the cremasteric muscle and increases the percentage of undescended testes

The child, finally in a cooperative mood, is examined first while standing Position of the testicle is determined, it may be in the perineal region, but usually lies somewhere in the inguinal canal If no testicle is palpable, the boy is asked to bear down, occasionally an intra-abdominal testicle may pop out into the inguinal canal At the same time the extent of the associated hernia is determined In our experience there has been an associated hernia of varying size in at least 90 per cent of cases of cryptorchidism Examination is continued in the supine position with repetition of the foregoing maneuvers Before making a statement that the testicle is missing (very rare) or that it is in the abdomen (moderately uncommon), it is well to go over the inguinal and perineal regions again If, as usual, one finds a testicle in the inguinal canal, repeated attempts are made to tease it into the scrotum

In those instances in which the testicle can be brought into the scrotum, it is wise, as the testicle is being held, to have the parents feel it and note how it snaps back into the canal when released The child has been brought in with a diagnosis of undescended testicle for which, most likely, operation has been advised To convince parents that in this instance operation is unnecessary, a considerable point is made of the fact that they themselves have felt the testicle in the scrotum Then follows this unequivocal statement A testicle which lies in the inguinal canal, but can be brought into the scrotum, will eventually descend spontaneously, before or certainly at puberty To maintain good relations with medical confreres, it is well to confirm to the parents the diagnosis

Peritoneal neck of sac (sutured)
Ext obl m
Incision in int obl m
Spermatic cord freed
Testis
Peritoneum lifted
Spermatic vessels freed retroperitoneally
Taped to thigh

mature hankerings The use of gonadotropic hormones except at or near puberty is unphysiological and therefore condemned.

Bilateral undescended testes present a problem somewhat different from that of unilateral nondescent It is sometimes difficult, and probably unwise, to talk parents out of operation until the child reaches ten to eleven years of age The almost invisible scrotum in these boys may be the cause of a psychologic problem when the boy goes to school, observes other boys, and is in turn observed and probably ridiculed The fact that torsion of the testis is more likely to affect an undescended testis than a normally placed one is also a factor to be taken into consideration We have no fixed rule for delaying operation until approaching puberty in boys with bilateral cryptorchidism If the parents are insistent, are worrying about the defect and have been strongly advised by their pediatrician or family physician to have early operation, we proceed, but not before the child is about six years of age unless, as stated above, large associated hernias require earlier operation We never operate on both sides at the same time, in fact, we usually space the operations at least one year apart to allow repeated observation of the result of the first operation

THE OPERATION. Trustworthy guides for successful placement of an undescended testicle in the scrotum are adequate incision, wide exposure, complete release of all impeding strands of tissue and, above all, thorough liberation of the spermatic blood vessels from surrounding structures

Under general anesthesia a long inguinal incision is made The external oblique muscle is split longitudinally along its fibers at such a level that the external ring is bisected The testicle, usually lying somewhere in the canal, is elevated along with all the structures of the cord and separated from its fibrous connection with the gubernaculum With a finger in the scrotum the dartos muscle is stretched to make room for the testicle The internal oblique muscle is cut transversely for about 1½ inches parallel with the inguinal ligament It is impossible to perform an adequate dissection without the added room obtained by cutting this muscle The hernial sac is easily identified and opened by a small longitudinal incision With two fine hemostats on each side, the hernial sac is laid open, held under tension, and separated from the vessels and the vas with fine embroidery scissors The distal end of the sac is disregarded, and the proximal end is completely freed from the vessels of the cord, transfixed and ligated high with a silk suture The abdominal peritoneum is retracted medially with a broad flat retractor as the vessels of the cord are freed proximally to the level of the kidney Painstakingly, all bits of fibrous tissue attached to the vessels are cut If the general rule of never cutting any tissue which, when held up with an instrument, is not clearly translucent is followed, there is little danger of injuring important structures The utmost care must be exercised in the dissection and

Peritoneal neck of sac (sutured)

Ext obl m

Incision in int obl m

Spermatic cord freed

Testis

Peritoneum lifted

Spermatic vessels freed retroperitoneally

Taped to thigh

intercostal nerves in the chest, and the balance from nerves in the pelvis and neck, and from unknown sources

Diagnosis

The child suspected of having a neuroblastoma is usually less than five years old, approximately one third are less than one year old Contrary to the minimal clinical findings associated with embryoma of the kidney, the child having a neuroblastoma is usually pale and sickly looking and has lost weight Not infrequently generalized metastases from a neuroblastoma, the origin of which sometimes cannot be determined, already have occurred when the child is first examined Because of the insidious onset of deep bone pain it is understandable why many of these children have been treated for arthritis Only that child is relatively free from symptoms in whom by good fortune the tumor has been discovered early

The tumor, lying fixed in the upper flank, is irregular in size and usually extends across the midline The positive shadow on the x-ray film of the abdomen outlines the size and position of the tumor In excretory urograms flattening of the kidney pelvis suggests neuroblastoma, whereas distortion of the kidney pelvis indicates a Wilms' tumor Deposits of calcium in the tumor are diagnostic of neuroblastoma Rarely is it impossible to differentiate a neuroblastoma from a Wilms' tumor

Whenever a tentative diagnosis of neuroblastoma has been made, x-ray films of the head are taken because the skull bones are usually the first site of metastases A lesion in the skull calls for complete roentgenographic bone survey In case a metastatic lesion is found in the skull and the rest of the skeleton is free from metastases, I believe the abdominal tumor should be removed in whole or in part because neuroblastomas behave unpredictably Obviously, operation is contraindicated if metastases are generalized

Treatment

The neuroblastoma arising from the adrenal is most satisfactorily approached through a long, subcostal, transverse abdominal incision It is, of course, desirable completely to remove the tumor In those instances in which this objective is unattainable because the new growth surrounds and is fixed to important structures, it is well to excise as much of the tumor as possible Instead of spreading cancer cells by such trauma, as is true in other cancer surgery, it actually appears at times that such manipulation is beneficial

After operation roentgen therapy is immediately begun The prognosis after complete removal of an abdominal neuroblastoma is fair, the

younger the child the better the prognosis Seven of the 8 cures in our series of 31 cases were in children below 1 year of age the youngest 6 weeks old at the time of initial treatment—a survival rate of 25 per cent When may a child be considered cured of neuroblastoma? I don t know Usually if no recurrence has appeared within eighteen months, the prognosis is good And yet one of our patients carried in the "cured" column for three years rather suddenly became acutely ill and within a few months died of generalized metastases The prognosis should be guarded because the course of a neuroblastoma is unpredictable. Two cases illustrate this point.

A ten month-old child was operated upon in 1946 because of a large retroperitoneal mass Surgical removal was impossible only a biopsy was done The tumor presumably a neuroblastoma, was made up of nondescript primitive cells many in the process of mitosis The parents were told that the outlook was bleak X ray therapy was given the tumor melted away and the child today thirteen years later is in perfect health.

A healthy appearing 6-month-old boy was admitted to the hospital because of a tumor about 4 cm in diameter and 1 cm thick on the back of his neck a similar mass was found beneath the skin in the right lower quadrant Calcium deposits in the nonpalpable retroperitoneal tumor were visible in the x ray films. The tumor on the back of the neck was excised and by histologic examination proved to be a neuroblastoma After irradiation the tumor in the right lower quadrant disappeared Within the next two months similar tumors in the region of the right thigh, left buttock and left hip appeared and were successfully irradiated Four months after the initial examination a swelling arose above the left eye X ray films confirmed the suspicion of bone metastases and x radiation was given. The mass disappeared, and the bone regained its normal texture. The boy now seven years old is well and apparently free from disease

Such good fortune in the treatment of neuroblastomas is rare Why a malignant tumor will completely disappear or transpose itself into a benign neurofibroma is unknown. Too often explosive metastases unresponsive to any treatment, are fatal within a few months after the diagnosis has been made.

A four year-old girl, riddled with metastases six months after removal of an abdominal neuroblastoma, died in our hospital. During the last few weeks of life she quietly and uncomplainingly lay in bed. So long as she wasn t moved she suffered little pain Her favorite toy was a musical teddy bear She loved to have anybody stop at her bedside and wind up the music box. Over and over again it played the plaintive strain of Brahms Lullaby Can anyone stand at the bedside of such a

child and not be stirred by compassion and stimulated to search for more effective treatment?

EMBRYOMA OF THE KIDNEY (WILMS' TUMOR)

Diagnosis

No practical method is known for making a diagnosis of embryoma of the kidney before the tumor becomes palpable. Either the mother notices enlargement of the abdomen, or the tumor is found by the pediatrician who makes it a habit to feel the abdomen of every child brought to his office, even though the ailment is nothing more serious than a running nose.

The tumor, often the size of an orange or grapefruit, lies in either flank, is smooth, round, firm and painless. Irregularity of the kidney pelvis outlined in excretory urograms makes a specific diagnosis of Wilms' tumor possible in most instances. An x-ray film of the chest must be taken before operation is advised because the lungs are usually the first site of distant metastases.

Treatment

Immediate surgical removal through an ample, subcostal transverse abdominal incision is the acceptable present-day treatment of embryoma of the kidney. (Lumbar incision is unconditionally condemned.) The splenic or hepatic flexure of the colon is freed and drawn medially, and with a minimum of trauma the hilus of the kidney is approached. The artery and vein are isolated, doubly ligated and cut. From this point on the operation consists merely in tying the ureter as far distal as possible and freeing the mass from the kidney recess. The entire operative area and the region along the aorta are carefully examined for lymph nodes, and all, regardless of size, are excised.

The skin wound is covered with a small dressing fixed to the skin with Scotch tape rather than adhesive tape because of the roentgen therapy which is to follow. Daily doses of irradiation to the appropriate flank are given until the child has received the maximum treatment. Most centers at which many embryomas of the kidney are treated—notably the Children's Medical Center in Boston—emphatically advise vigorous irradiation of the operative site beginning immediately after operation. For the past twelve years we have followed this regimen, and yet we can't help questioning the justification of concentrating so much radiation upon an area from which a well encapsulated tumor has been removed without apparent spillage of cells. The fact that later metastases may appear in the lungs rather than in the abdomen suggests that a few

heavy doses of x ray therapy to the lungs immediately after operation might be just as beneficial as radiotherapy of the abdomen or might be given in addition to radiation of the abdomen Prophylactic postoperative roentgen therapy to the lungs is not advised—it is only suggested for consideration

Preoperative x ray therapy to shrink an unusually large tumor has been advised by some to simplify later operation The necessary delay of four to six weeks while awaiting the full effect of irradiation hardly seems justifiable Every day operation is delayed the danger of me tastasis is increased Wilms tumors are not irremovable because of their size—only because of local extension of the growth.

The question repeatedly has been asked Shall isolated metastatic nodules appearing in the lung after excision of a Wilms tumor be re moved surgically or be irradiated? If the nodule or nodules appear to be confined to one lobe surgical excision I believe, is preferable. Other tiny nodules invisible in the x ray film may be scattered throughout the lungs Nevertheless, the long chance of removing the only metastatic growth in the lungs seems to justify the operation

Combined operation and postoperative x ray therapy at our hospital have resulted in approximately 30 per cent cures this figure has been constant during the past 12 years

There seems at present to be little hope for the child who has local recurrence of a Wilms tumor and/or widespread metastases A second series of x ray treatments to the site of painful metastases sometimes is beneficial for a short time.

During recent years intravenous nitrogen mustard has been fairly extensively used for the treatment of generalized recurrences of Wilms tumors, but apparently with little if any success It may be that occa sionally nitrogen mustard prolongs life a few weeks The fact that the drug makes some children rather ill for a number of days and that long term benefit has not followed makes it questionable whether this drug should be continued for the treatment of metastatic lesions The child who in spite of widespread metastases is happy and still has a good appetite will, I believe receive just as much benefit from a nice, juicy nitrogen rich hot dog touched with mustard to taste and taken by mouth as from nitrogen mustard injected into a vein. Children much prefer the hot dogs Maybe the temporary happiness of a satisfactory meal is worth as much as a few later days of just living Relentless search for an effective cancerocide must go on.

CYSTIC HYGROMA

Cystic hygromas or cavernous lymphangiomas are benign tumors and remain so These loculated cystic masses occasionally appear in the

axilla or groin, rarely in the chest or abdomen, but commonly in the region of the neck, where they may extend in all directions around nerves, blood vessels, the trachea and esophagus and into the muscles. A cystic hygroma, although benign, completely disregards lines of cleavage.

Diagnosis

An unusually soft, nontender, fluctuant, cystic swelling present at birth or appearing shortly thereafter, located in either the anterior or posterior triangles of the neck or both, is so characteristic of cystic hygroma that diagnosis is rarely a problem. The mass varies in size from as large as the child's head to a barely visible nodule. Hemangiomatous elements in the cystic mass are usually recognizable by the bluish tinge they give to the skin. A firm mass suddenly appearing in a child's neck suggests hemorrhage into a cystic hygroma not previously recognized. Infection in the untampered-with cystic hygroma is rare.

Treatment

Surgical excision is the treatment of choice. This statement, however, requires some modification. If the swelling is barely visible, no treatment is necessary, provided the parents can be convinced of this fact. Occasional observation suffices; if the mass increases in size, and excision becomes advisable, nothing has been lost by waiting. Commonly the swelling does not change in size and, as the child grows, becomes less apparent.

X-ray therapy has been unjustifiably condemned as useless. 'Tis true, it often fails and should not be used on large swellings, but it is worthy of trial in those cases in which a moderate-sized, rather flat mass is situated below the ear and spreads out over the parotid region. In this area lies the facial nerve. I confess that I am more afraid of injuring the facial nerve than any other structure encountered in surgery. Because of this cowardice roentgen therapy is requested, and a series of 3 treatments of 400 roentgens each is given. The full effect of irradiation is not apparent for three or four months. If then there has been some improvement, the course of irradiation is repeated. After x-ray therapy the tumor has occasionally shrunk sufficiently that parents are satisfied with the cosmetic result. The fact that remnants of the cystic hygroma remain worries me far less than the mental picture of a partially paralyzed face. In the surgical treatment of approximately ninety cases we have erred at times in being too bold or too timid. Memory of errors persists far longer than that of successes. The proper attitude to take toward surgery of cystic hygroma is that the operation will never be simple.

Regardless of the size or shape of the mass a transverse incision in

the skin along the lines of cleavage is important for a good cosmetic result. The most superficial portion of the cyst is first exposed So far as possible it is well to leave the cyst or cysts (they usually are multiple) intact and dissect them from surrounding structures A unilocular or multilocular cyst not extending about important structures is rare. Removal of a cystic hygroma in the posterior triangle of the neck is simple However masses overlying the sternocleidomastoid muscle and extending forward in the anterior triangle of the neck and along the inferior surface of the ramus of the mandible call for caution After the superficial portion of the mass has been isolated smaller cysts will be found and there is no limit to the possible extent of these cysts The location of the spinal accessory the recurrent laryngeal and the superior laryngeal nerves must always be borne in mind and the fact that they may be pushed into abnormal locations must not be forgotten The vagus nerve the carotid artery and jugular vein are easily avoided

An overzealous attempt to excise all of a cystic hygroma extending from the floor of the mouth to the apex of the lung and surrounding the trachea and esophagus led to distressing postoperative problems in a child upon whom I operated After operation a tracheotomy was necessary to relieve dyspnea The child was unable to swallow without choking and had to be fed by gavage The recurrent laryngeal and most likely the superior laryngeal nerves had been injured For weeks the care of this child was a twenty four hour a-day problem in feeding, maintaining an adequate airway and in preventing aspiration The child eventually recovered, but memory of the operative and postoperative problems remains undimmed To get a good result not all the tiny cysts need be removed it is sufficient to cut the tops from those which lie in danger areas

The objectives in surgery of cystic hygroma are relief of obstruction upon vital structures and a good cosmetic result. Good judgment must control the extent of the operation. Inadequate operation is just as inexcusable as too daring operation, and nowhere does this maxim apply more forcefully than in the surgical treatment of large cystic hygromas of the neck.

SACROCOCCYGEAL TERATOMAS

Teratomas arising from the sacrococcygeal region are presumably remnants of tissue intended to be a twin. The tumors vary in size from a small nodule to one third the size of the baby are readily diagnosed and should be removed in the immediate neonatal period Although the majority of these tumors are largely external a few extend proximally and may fill the pelvis

Some of these tumors—about 85 per cent—are benign, and the bal

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Postscript

This book is not finished—I've just stopped writing Only some of the conditions common to pediatric surgery have been briefly considered Lest anyone get the idea that children's surgery is limited to so few subjects I hasten to add that a book many times this size could not adequately cover the surgical problems encountered from year to year So much of infants surgery is unusual. Variations of common congenital anomalies are the rule rather than the exception A detailed chapter on atresia of the rectum and all its complications would require a volume half this size

Many dictatorial statements have been made deliberately to sharpen the focus upon diagnosis and treatment as they are seen in the light of today's knowledge Tomorrow many of these pronouncements will be incorrect as new discoveries replace established truths. Pediatric surgery is a new and rapidly changing specialty Better methods of diagnosis and improved techniques of surgery will soon make obsolete much that has been here written.

The ultimate objective remains the same to give the deformed or surgically ill infant and child the best care available. Their misfortune is our problem To them this book is rededicated.

ance are malignant. It is doubtful whether a benign tumor transforms itself into malignant form It appears that at birth a sacrococcygeal tumor already is either benign or malignant This assumption is based upon examination of tumors which had been partially removed shortly after birth, and completely removed or biopsied a year or two later The microscopic findings did not change, all were either benign or malignant from the beginning

Careful examination of the child and appraisal of possible technical difficulties is in order before sending the child to the operating room Twice as much blood as seems necessary is provided for transfusion because bleeding is apt to be brisk In fact, most of the children upon whom we have reoperated were those in whom the previous surgeon had stopped the operation short of complete removal because of shock

The child is placed on the operating table in the prone position, the buttocks are elevated with a sandbag or pillow After surgical preparation of the area a large sterile catheter or rectal tube is inserted into the rectum It is reassuring to feel the tube from time to time as the tumor is being dissected from its intimate attachment to the rectum

The coccyx is always removed It is desirable that the tumor be removed en masse without fragmentation because it may be malignant Occasionally after having freed the posterior segment of the tumor it is found that an intrapelvic mass cannot be reached If thus far all has gone well, the baby may be turned over and the pelvic mass freed through an abdominal incision

A malignant teratoma fixed in the sacrococcygeal region or in the pelvis, and not removable without injuring the rectum and bladder, should be left alone Even if it were possible, I would not attempt a pelvic exenteration in a newborn child

The subject of tumors in children has barely been touched

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